

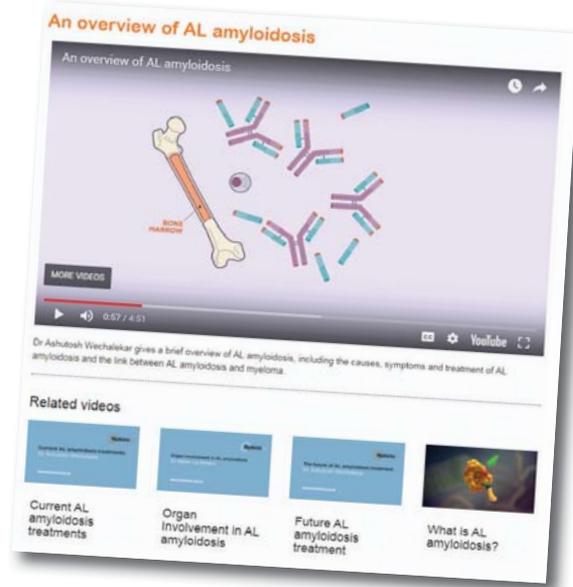
# AL amyloidosis Matters

Issue 15 Autumn 2017

## New patient videos available from Myeloma UK

Myeloma UK has recently added to its range of AL amyloidosis information with the production of four videos which are hosted on our website.

The video titles are: An overview of AL amyloidosis; Current AL amyloidosis treatments; Organ involvement in AL amyloidosis; and Future AL amyloidosis treatment. They feature Dr Ashu Wechalekar and Dr Helen Lachman from the National Amyloidosis Centre and aim to help patients and their families understand more about the disease.



You can view the videos by visiting the AL amyloidosis 'channel' at: [www.myeloma.org.uk/mtv](http://www.myeloma.org.uk/mtv)

Written information about AL amyloidosis can be downloaded or ordered hard-copy through the Myeloma UK website at: [www.myeloma.org.uk/publications](http://www.myeloma.org.uk/publications)

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## Welcome

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

In this edition you can read the final instalment of patient Richard Thompson's book - Meeting Amy - and articles on managing bowel problems and writing a Will.

In our patient experience feature, Elizabeth Doy shares her experience of living with AL amyloidosis for the past 9 years.

And finally, don't forget our annual AL amyloidosis Infoday is taking place on Friday 17 November in London - to book a place call us on 0131 557 3332.



Jude Leitch  
Editor

## AL amyloidosis Matters

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

# News & Notes

## Study shows daratumumab to be highly active in AL amyloidosis

In a study of 25 relapsed AL amyloidosis patients, daratumumab (Darzalex®) has been found to be highly effective. Daratumumab is a monoclonal antibody drug licensed for the treatment of myeloma. The overall haematological response rate (a treatment response determined by the reduction of free light chains) to daratumumab was 76%, including a complete response

in 36% and very good partial response in 24% of patients. The treatment was well tolerated, even among the 72% of patients with advanced cardiac involvement. Although based on a small study group, the study provides evidence for the activity of daratumumab against AL amyloidosis and compares favourably with other existing treatment options in the setting of relapsed AL amyloidosis.



## Ixazomib shows promise in AL amyloidosis

The first study to assess the oral proteasome inhibitor drug, ixazomib (Ninlaro®), in relapsed and/or refractory AL amyloidosis patients has been published. In the Phase I/II trial of 27 patients, ixazomib provided an overall response rate of 52%, and the drug was well-tolerated. This is encouraging as there are challenges involved with administering the other proteasome inhibitors - bortezomib (Velcade®) and

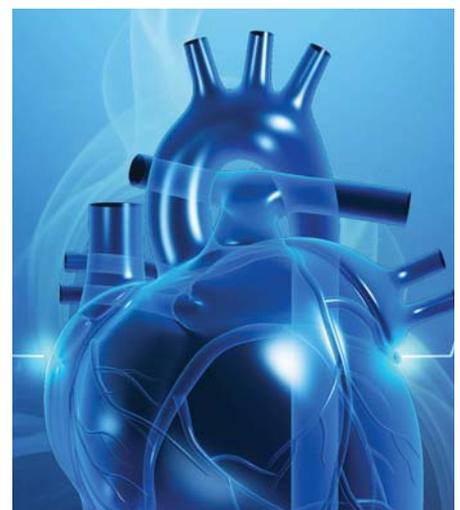
carfilzomib (Kyprolis®) - to AL amyloidosis patients because of the increased risk of neuropathy (including autonomic neuropathy) and heart-related side-effects, respectively. Importantly, peripheral neuropathy observed within the trial participants was mild and infrequent, and therefore ixazomib could be a more appropriate treatment option for patients who have neuropathy. Ixazomib is now being studied in a number of Phase III trials in AL amyloidosis patients.



## New research supports clinical relevance of cardiac biomarker NT-proBNP in AL amyloidosis

Research presented at a recent conference focused on heart failure further supports the important role of the cardiac biomarker NT-proBNP in the biology of AL amyloidosis. New preclinical research shows how misfolded light chains, the hallmark of AL amyloidosis, induce toxicity to heart cells and increase the production of NT-proBNP. This supports the

relationship that has previously been reported between lower NT-proBNP levels and improved cardiac function and outcomes in AL amyloidosis patients. "These results provide new insights into the important role of NT-proBNP in AL amyloidosis and offer further support for the clinical utility of NT-proBNP as a surrogate biomarker in AL amyloidosis studies" said the lead researcher.



# Meeting Amy

## - a cautionary tale (PART THREE)

BY RICHARD THOMPSON

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

### June 2016

You know the worst thing about being given a get-out-of-jail-card? I feel a fraud.

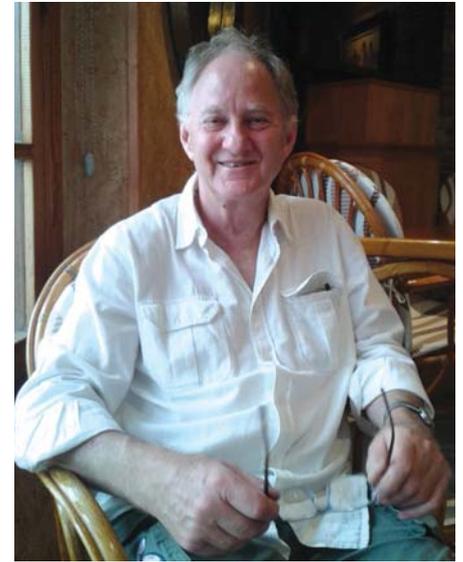
It's the thinness of the veil that separates life and death that's so bewildering. One minute you are here, the next you are gone. Being *ill* really makes you think about this stuff. But you learn to cope through the strength of others. For now, at least, I can say I got off lightly, because I'm *in remission*. It's a frigging miracle. Even my consultants seem surprised.

A few weeks back I went through a totally unexpected - and uncharacteristic - bout of post-chemo blues. You're not prepared for a serious downer because, on the one hand you don't see how you could feel depressed about getting better, and on the other, you don't think you have any right to feel sorry for yourself. Double whammy. The truth is that all the time you are focussed on just getting through the chemo, you don't have the time to worry how you feel about it. When it comes to an end and you start to recover from the treatment (not the illness), you suddenly find yourself with too much time on your hands. Time to ponder the 'what if's' and

'why me's?' And because you feel so much better *physically*, you think you're being rational. But you're not. Your emotions are all over the place. However brave people said you were, you know in your heart of hearts that every day was a fight to stay on top, not just for yourself, but for those you love too.

Then the survivor guilt thing kicks in. What right have I to still be here when those lovely kids I saw on Chemo Suite might not be?

I've just come back from my six-monthly super-check at the National Amyloidosis Centre in London and they're very pleased with my progress. I can live a more or less normal life again and they'll continue monitoring me to make sure I'm not slipping backwards. To be honest, I don't know what comes next and I don't particularly care, providing I stay healthy and positive. There's nothing like a shock to keep you focussed. I always thought I was invincible. Now I'm just grateful I'm still here. It could have been so much worse, but it wasn't. I know people who have undergone horrendous treatment for conditions far worse than mine and have huge respect for their resilience. You see kids with drips going into their arms and you'd do anything to swap with them because it seems so unfair



their young lives are blighted by ill-health. Yet they don't make a fuss. They just get on with it.

Then there are those of you who did everything you could for someone you love, yet still lost them to eternity. That *ache* is beyond comprehension unless you've been through it. I'm raising a glass to all my friends, both present and absent. Thank you. You've been sensational. And I'm raising another to those fantastic doctors, nurses and support teams who've made me better and given me back my future.

*I'm lost for words.*

### Post-script - September 2017

Gazing at the azure blue sea in Asturias, northern Spain, I realize how privileged - not lucky - I have been over the past couple of years in my treatment and care both in London and Leicester. I'm fine now. At 71 I feel good and even if my kappa light chains go up and down a bit, I know I'm in good hands. When I was first diagnosed I thought I was on the way out. Today I'm most certainly back in and I intend to stick around for a good few years yet.

Meeting Amy wasn't so bad after all.

# Tips for managing bowel problems

BY ALICE BARON  
Patient Information Officer, Myeloma UK

**Bowel problems like constipation and diarrhoea are common in AL amyloidosis. They can leave you feeling fatigued, uncomfortable, with stomach cramps and a reduced appetite. It's important to discuss them with your doctor or nurse, even if you feel embarrassed, so they can help you manage them.**



## Constipation

Constipation is usually diagnosed when your bowels are moving fewer than three times a week, and any movements you do have may be dry, hard and painful.

There are a variety of causes of constipation in AL amyloidosis. Amyloid deposits along the gastrointestinal (GI) tract or along the nerves that control the muscles of the tract can affect the normal digestive process and cause constipation.

Constipation can also be a side-effect of some AL amyloidosis treatments; caused by lifestyle factors like poor diet, dehydration or lack of exercise; or caused by psychological difficulties like anxiety, depression and stress.

### Tips for managing constipation:

- Adjust your diet to include more fibre – eat more vegetables, dried fruit and wholegrain carbohydrates
- Eat small meals frequently throughout the day
- Stay well hydrated but avoid caffeinated drinks
- Try natural remedies such as fibre supplements, linseed oil or syrup of figs. Speak to your doctor before trying a

supplement to make sure they don't interfere with your AL amyloidosis treatment

- Exercise gently and regularly
- Try anxiety and stress relieving techniques
- Discuss taking laxatives with your doctor

**“TRY TO ADJUST YOUR DIET TO MAKE SURE YOU'RE GETTING ENOUGH FRUIT, VEG AND NUTS. I BOUGHT A JUICER.”**

## Diarrhoea

Diarrhoea is the passing of loose or watery stools more than three times a day and can make you dehydrated if left unmanaged.

Like constipation, diarrhoea can be caused by amyloid deposits in and around the GI tract, anxiety, stress and some AL amyloidosis treatments. Antibiotics and infections can also cause diarrhoea.

Diarrhoea can be particularly distressing because the unpredictability and urgency of it may restrict what you feel able to do.

- Stay well hydrated but avoid caffeinated drinks

- Eat small, frequent meals throughout the day
- Avoid spicy or fatty foods
- Use soft toilet paper or moist wipes
- Wear comfortable clothes and rest when necessary
- Maintain good hygiene
- Discuss taking over-the-counter treatments with your doctor

AL amyloidosis patients need to control their salt and fluid intake to prevent fluid overload, so speak to your doctor or nurse about whether any limits need to be adjusted to replace the salts and fluid lost through diarrhoea.

## Summary

Bowel problems are common in AL amyloidosis and should be discussed with your doctor or nurse. You may be prescribed treatment for your bowel problems but you can help manage them yourself, particularly by paying close attention to what you eat and drink.

For more information see the *AL amyloidosis Constipation and Diarrhoea* Infosheets from Myeloma UK.

## Writing a Will

BY SELENA HALLAHAN  
Patient Information Officer, Myeloma UK

**A Will is a legal document in which a person leaves instructions about what they want to happen to their property, money and possessions (also known as an estate) after their death. Most people make a Will to ensure that their estate goes to the right people.**



### Writing a Will

It is a good idea to contact a solicitor who can help to write a Will and ensure that nothing is left out.

Some people prefer to write their own Will to avoid solicitor fees, however it's important to bear in mind that the slightest error or misuse of legal terms can invalidate the Will and may prevent a person's wishes from being carried out after their death.

Every November is National Will Aid month. Will Aid is a partnership between the legal profession and nine charities. In November, participating solicitors waive their fee for writing a basic Will and invite clients to make a donation to charity instead.

For more information go to [www.willaid.org.uk](http://www.willaid.org.uk)

### Things to consider when writing a Will

Before writing a Will, it helps to think about the following:

- The property, money and possessions that make up the estate
- How the estate should be distributed and to whom
- Any specific funeral arrangements

- Who to appoint as legal guardians if there are children under 18
- The executor(s) of the Will – this is someone who is chosen by the person writing the Will who they trust to deal with their estate according to the wishes stated in the Will once they die. Executor(s) can be relatives, friends or a professional but before naming them in the Will, it is important to check that they are happy to take on the role

### Signing a Will

To ensure a Will is valid, it needs to be signed by the person writing the Will, in front of two witnesses who also have to sign it.

Witnesses are required to be people who will not benefit from and who are not executors of the Will. To avoid problems or conflicts of interest most solicitors use staff in their offices to act as witnesses.

### Who looks after a Will?

A Will needs to be stored safely at home or lodged with a solicitor or bank.

Normally when a solicitor makes a Will, they keep the original and

a copy is given to the person writing the Will. It is important to let the executor know where the Will is stored. It is also important for an individual to maintain their Will every few years or after any changes in life circumstances, for example:

- Separating from a partner or divorcing a spouse
- Getting married (this cancels any previous Will)
- Having a child or grandchildren
- Moving house
- If the named executor(s) dies

### Summary

Writing a Will is important in ensuring a person's estate goes to those they want it to, no matter how big or small the item(s).

Getting your affairs in order before you die, by making a Will or planning your funeral, will help make the practical tasks much simpler for your family members during a difficult and emotional time.

A simple Will is not usually expensive and could save families legal fees and disputes in the future.

# PATIENT EXPERIENCE

**Elizabeth Doy is 62 and lives in Leicestershire with her partner. She was diagnosed with myeloma in April 2008 and then AL amyloidosis after a kidney biopsy in September that year.**

**I am pleased to be offered the opportunity to share my story, but with so many facets to it the difficulty is what to concentrate on. Where to start?**

I had worked in a very busy pharmacy for fifteen years until a few months before my diagnosis. The job had become utterly exhausting and I made the decision to leave and work in a general store a short walk from home. Soon the first physical symptom became apparent - my urine turned frothy. A visit to my doctor confirmed my suspicion that it was protein. Blood tests indicated myeloma, and probably amyloid involvement in the kidney, causing albumin to be leaked from the bloodstream into the urine.

I was relaxing after work on a Friday evening when I received a call from my doctor with the news. My world seemed to turn on its head. Not having a computer at the time I dug out my ancient medical encyclopaedia which informed me I wasn't likely to live more than five years. My long term partner didn't seem to be able to deal with the news and went away for the weekend, leaving me to contemplate the future. There was no family to tell besides my elderly mother as I have no children or brothers and sisters.

I tried to start out upbeat and positive, but the situation rapidly became increasingly difficult to deal with. From being what I would like to describe as a coping, rational and organised person, I became the exact opposite. After being initially sympathetic my partner grew tired and intolerant of my mental state and threatened to leave if I didn't improve, which of course made me worse. He did eventually leave after some months - an awful time that is difficult for me to even think about.

Initial treatment was the combination of dexamethasone, cyclophosphamide and thalidomide. I only had a partial response and moved on to being treated with bortezomib (Velcade®) combined with dexamethasone, again with only a partial response. I think that these results were because I was not taking the dexamethasone consistently because of my mental state. I can't begin to explain how I felt, but I suppose I was having some kind of mental breakdown.

It was at this time, when I was in the daycare unit having an infusion of bortezomib, that a nurse realised I needed psychiatric help and it was arranged for me to see a psychiatrist who specialised in cancer patients. A member of his team visited me at home every three weeks for over a year,

and I am indebted to them for eventually getting me back to normal.

Financial problems were there from the start. I stopped working when I started chemotherapy - oedema (swelling) for example, had become a significant problem because of my kidney function. I also felt mentally and physically ill. I had only been at my new place of work for a few months so could only have statutory sick pay. I was incredibly worried that I would get into arrears with my mortgage. Thankfully a good friend arranged for me to see an advisor from the citizens' advice bureau who helped me to claim the benefits that I was entitled to.

Following the course of bortezomib, I had a week's course of a combination chemotherapy in hospital followed by stem cell harvesting. This was now May 2009, and in the July I had high-dose therapy and a stem cell transplant (HDT-SCT). Throughout all of this time my mental state was awful and life because of this was a waking nightmare. I was crippled by anxiety, couldn't talk rationally and was showing signs of agoraphobia. I wondered if I would ever be normal again.

I convalesced at my mother's post HDT-SCT. This had been insisted on by my GP and haematologist because of the fact I now lived

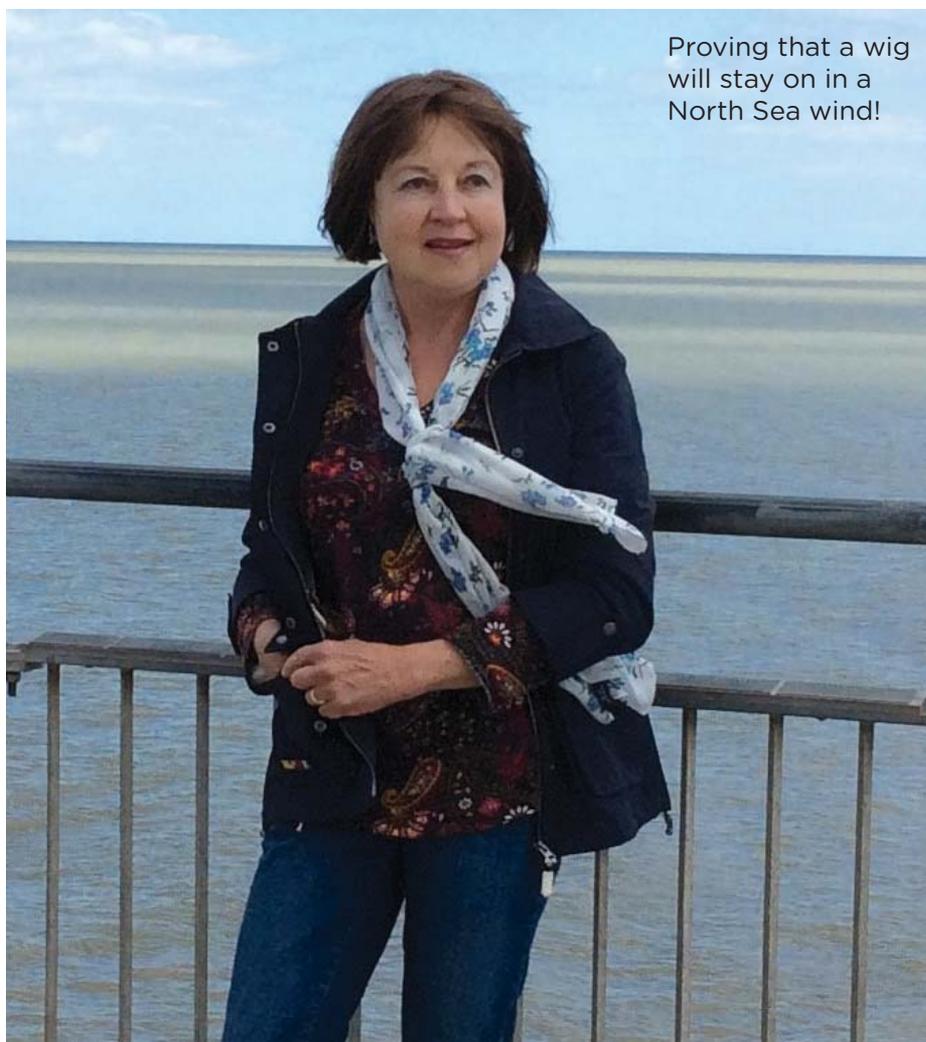
alone. Unfortunately, my kidney function deteriorated to the point when I needed emergency dialysis in early January 2010.

When I first started dialysis I was very ill indeed, but within six weeks I was starting to feel better. Amazingly this coincided with an improvement in my mental state after I had been prescribed an antidepressant combined with an anti-anxiety drug.

By April 2010 I was living back at my own home. Dialysis proved to be little of what I had been dreading. It rapidly became just another part of my life, I made many friends - all of us in the same boat so we were able to joke about our predicament. The staff were lovely even though the unit was extremely busy.

I have been lucky that my myeloma has been in partial remission for so long. I produce light chains but their output until now has been stable, as has the low level of paraprotein (the myeloma marker) that I am producing. I have over the years lost my hair and nails. Salivary gland involvement means I have swellings in my neck and a dry mouth. Recent tests indicate I may have some minor cardiac involvement, and I realise at some point I will need further treatment. This is something I can now face with confidence, and I have learnt for a long time that this a disease I am living with and not to ruin the present by worrying about the future. I am able to arrange a meeting with a medical psychologist who specialises in renal patients if the situation ever starts to get me down, something I wouldn't hesitate to do.

So what of my life now? I paid off the mortgage in 2012 - all my initial anxieties were for nothing! After three years of living happily alone I met up again with a former work colleague who knew all about my medical condition.



Proving that a wig will stay on in a North Sea wind!

Within a few months we decided to live together. Because of my new circumstances I realised that I could be a candidate for a home dialysis machine, something that is encouraged where possible. I trained to operate the machine and learnt how to put my own needles in (I have haemodialysis), and have for the past three years dialysed at home.

My partner is very supportive and unfazed by my health problems, with very much a 'live for the day' philosophy. Life is good and I work the dialysis round it, an advantage of having a home machine. I volunteer at a fairtrade shop, where I can do as much or as little as I feel able to. Visits to the National Amyloidosis Centre in London are at the moment annual and a good opportunity to have

a shop in Hampstead!

Over these years Myeloma UK has been at the end of the phone for any questions. I recommend their Infodays if they're possible to get to. My treatment and care from the word go have been excellent and the only thing that I think could have been done differently is an earlier referral to a psychiatrist as it was very obvious I was experiencing major problems.

This story is really an overview of the past few years, and I've not detailed a lot of the things that have happened in order to fit it all in. Biopsies have not been mentioned, of which there have been several, and the dialysis angle is a whole different story, but I hope that this account will be of interest and proof that there is a life to live when AL amyloidosis becomes part of it.



## Host a Coffee Morning this November

Share a cuppa and a cookie with your family, friends, colleagues or community.

Hosting a **Coffee Morning** is a fun and easy way to **raise funds and awareness** and our **Fundraising Team** can help with tips and advice.

To find out more visit  
[www.myeloma.org.uk/coffee](http://www.myeloma.org.uk/coffee)  
or call **0131 557 3332**

## MAKE A DIFFERENCE THIS CHRISTMAS

Myeloma UK works in partnership with the National Amyloidosis Centre to fund research and provide information and support to patients and their families.

### You can support our work:



#### Donate instead of gifts

Ask your family and friends to donate to Myeloma UK instead of buying you gifts. Or donate to Myeloma UK instead of giving gifts.

Donate via our website  
[www.myeloma.org.uk/donate](http://www.myeloma.org.uk/donate)



Give as you Live™

#### Shop online

Shop online through Give as you Live™ and a percentage of every pound spent will be donated to Myeloma UK, at no cost to you.

Sign in to Give as you Live™ before you shop online  
[www.myeloma.org.uk/giveasyoulive](http://www.myeloma.org.uk/giveasyoulive)



#### Festive fundraising

Organise a **Christmas concert** or **fair**, **cake** or **craft sale**, a **Christmas Jumper Party** or a **collection** at a local festive event.

Or burn off those holiday calories with a **Santa Run**, **Boxing Day Dip** or make a **New Year's Resolution**, all while raising funds for Myeloma UK.

Visit [www.myeloma.org.uk/christmas](http://www.myeloma.org.uk/christmas) for more information or call **0131 557 3332** or email [fundraising@myeloma.org.uk](mailto:fundraising@myeloma.org.uk)

**Thank you for your support.**

