Thalassaemia Matters

AUTUMN 2019
ISSUE 131

In this issue...

Transition
How to cope with periods of change

Infected Blood Inquiry
An interview with Jennifer Cole

National Thalassaemia Day
Screen your genes

...and much more!

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What is thalassaemia?

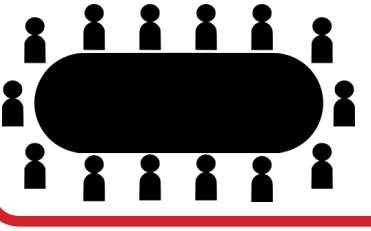
Thalassaemia major is a serious, genetic blood disorder. People with the condition cannot make enough haemoglobin in their red blood cells and are dependent on regular blood transfusions throughout their lives. If left untreated, a child born with thalassaemia would die in early infancy. Thalassaemia developed as an evolutionary response to malaria which is why it mainly affects those coming from regions such as Asia and South East Asia, the Mediterranean, South America, the Caribbean, Northern and Central Africa and the Middle East. Whilst it was initially prevalent in these regions, due to the migration of communities over the centuries, anyone can be at risk. In fact, every year 75,000 babies are born with thalassaemia major worldwide. With better education of those at risk and more effective screening, this inherited condition could be eradicated.

Produces specialist educational material for patients and healthcare professionals

Provides counselling, support and information to patients and families

What does the UKTS do?

Organises national events including medical conferences, training days and support groups



Raises funds for vital medical research and life-saving equipment

Thalassaemia Matters is produced (content, interviews and design) by Neelam Dongha. If you would like to give feedback or make suggestions, please email neelam@ukts.org.

Contents

Message from the Chair

Autumn has descended upon us and I'm wondering where the last nine months have disappeared to?
I would like to especially thank everyone for the positive feedback received for our magazine. I am also delighted to welcome our new members and subscribers to the Society and am happy to announce that membership is now free.
I do hope that you enjoy this edition and ask that you continue to submit your suggestions and feedback.

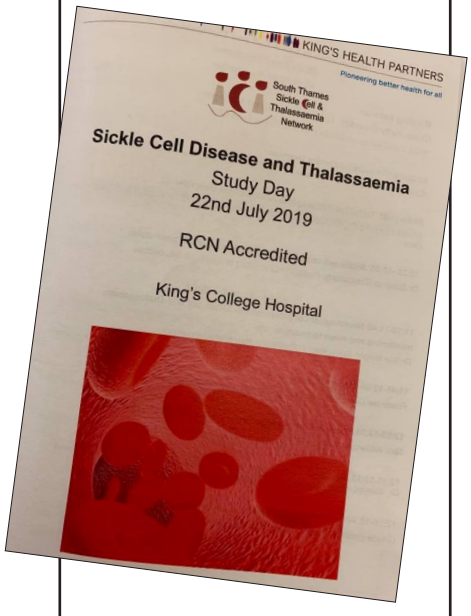
I would also like to ask everyone to help us publicise our National Day event on Saturday 19 October 2019. We are hoping to paint Southgate and the surrounding areas RED!

Please also help us with our fundraising efforts by supporting the Festive Charity Gala on Saturday 7 December 2019. Oddy, my fellow trustee, has been working very hard with her team to ensure that this night will be one to remember.
Looking forward to seeing you all at both events.

Gabriel Theophanous

Please follow the link to donate to us:
https://www.paypal.me/teamukts

KCH Study Day



We were invited to speak at the 3rd Annual Sickle Cell and Thalassaemia Study Day at Kings College Hospital run by Giselle Padmore-Payne and her team. The Study Day aims to educate health care professionals about these conditions.

Photo: Giselle Padmore-Payne and Roanna Maharaj (UKTS patient advocate)

MARIA FLETCHER TESTIFIES AT THE INFECTED BLOOD INQUIRY

In June Maria Fletcher testified at the Infected Blood Inquiry. Maria shared her experience of how receiving infected blood affected her life both medically and psychologically and how she was treated after receiving her diagnosis. We stand with Maria for her bravery and we hope the people responsible are held accountable for their actions and the impact on the many lives they have had. (See update on page 32).

Photo: Maria Fletcher and Andrew Credginton (UKTS)

UKTS Chair, Gabriel Theophanous, was interviewed for a new documentary on thalassaemia that is currently in the making!

Awareness-raising in Romford

Rachel Brown, Acute Clinical Nurse Specialist in Haemoglobinopathies at Queen's Hospital in Romford, invited us to go and participate in their awareness event. It was a great opportunity to increase understanding of thalassaemia.

Photo: Katerina Loizi (UKTS) and Rachel Brown

Photo: Dr Abimbola Momoh-Ojewuyi (Substantive Consultant and Romaine Maharaj (UKTS)

Photo: Dr Farah Barakat, Dr Gaby Corry, Dr Temuert Da Costa Gomez and Dr Florence Li

Thalassaemia event in Leicester

The UKTS participated in the East Midlands Sickle Cell and Thalassaemia Network (EMSTN) Patient and Family Education and Engagement Event in Leicester. The event was arranged by Rebecca Sekyere (Lead Nurse), Dr Amy Webster (Consultant Haematologist) and their colleagues across the network. Dr. Webster gave a short talk about the changes being made by the NHS with regards to commissioning and how it would affect patients. Professor Simon Dyson, a guest speaker, gave a talk about coping with sickle cell in schools. Rebecca Sekyere gave a presentation on how to treat and manage thalassaemia and sickle cell disease.

MAYOR PICKS THE UKTS!

On 24 June 2019, Romaine Maharaj (Executive Director) Andrew Credgington (Operations Manager) and Roanna Maharaj (Patient Advocate) met with Counsellor Kate Anolue, Mayor of Enfield at the Mayors Parlour in Enfield. The UKTS team gave a short presentation on the Society's history and the current work being undertaken. They also highlighted the challenges facing people living with or affected by thalassaemia. The Mayor whose professional career was in nursing and midwifery for forty years at the North Middlesex Hospital, understood how important it was to educate the public about thalassaemia. As a result of the presentation, the Mayor decided to add the UKTS to her list of chosen charities. She was surprised by how many communities were affected by thalassaemia.
We look forward to working with the mayor!

WELCOMING OUR GUYANESE VISITORS

We were delighted to be paid a visit by nurse Roxanne Siland and Dr Jewel Mahase, all the way from Guyana. We got a chance to discuss thalassaemia and the differing health systems with them, and we plan to work together to improve services for patients.

Photo: Roxanne Siland, Roanna Maharaj (UKTS), Dr Jewel Mahase

Welcome Adil Ray, OBE

The UKTS is delighted to introduce its newest patron, Adil Ray, who joined the team in July this year. Adil Ray OBE is best known as the star and creator of the hit BBC1 sitcom Citizen Khan. Five series have aired in the UK and the show is a global success. To date, Adil has received five Royal Television Society Awards for his work, including Best Comedy Programme and Best Comedy Performance as well as Best TV Character at the Asian Media Awards. In 2016, Adil was honoured in the Queen's Birthday List and awarded an OBE for services to media and broadcasting. Adil can currently be seen presenting Good Morning Britain for ITV. More on his work can be found via https://www.independent-360.com/presenters/adil-ray/.

Adil is happy to support the Society in its future work towards educating the public and improving treatment for patients living with thalassaemia. Upon his appointment, Adil tweeted the following message: "Pleased to help this amazing charity that aims to create awareness about a condition very few of us know about. Please do take a read when you have time." Gabriel Theophanous, Chair of the UKTS, said: "The Society is extremely grateful to Adil for his willingness to join the cause. The board of trustees and staff look forward to working with Adil in the future".

MEDIA COVERAGE:

The appointment of Adil generated a lot of media coverage and it was reported in Parikiaki (the Greek newspaper), Asian Today, Urdu Times and on CNN News TV.

Meetings attended by the UKTS (June-August 2019)

7 June
- Infected Blood Inquiry: Maria Fletcher's Hearing
- Blood Donation Campaign Forum

10 June
- UK Forum Meeting

18 June
Patrons' Meeting
-

21 June
- Brent Sickle Cell and Thalasssaemia Study Day

22 June
- Haemoglobinopathy Education Forum Leicestershire and South East of Leicester Sickle Cell and Thalassaemia Service

24 June
- Meeting with the Mayor of Enfield

8 July
- Screening Advisory Board Meeting

22 July
- Kings College Study Day

26 July
- Haemoglobinopathy Awareness Day Romford Hospital
- Visit from Guyanese team

30 July
- Meeting with Cellgene

18 August
- Leicester Belgrave Mela

22 August
- SCT Advisory Board Group Meeting

Various dates in August
- Training and hosting nurses from Nepal

Catching up with Dr Lakhani

It was a treat to have a familiar and friendly face pop in to the office whilst he was on a trip to the UK. Many of you will remember Dr Nitin Lakhani well from his highly successful time as President of the Society but what you may not know is that he now lives in Mauritius, is a qualified pilot and has three tortoises! We had a really interesting chat with him about his time at UKTS and all things thalassaemia and you can watch the full interview on our YouTube channel.

Can you briefly describe your employment history?

Before I became involved with the UKTS, I trained as a General Practitioner (GP). I qualified in Medicine in 1979 having started at Medical School in Birmingham in 1974, where I also met my wife Kamlesh! At some point during my medical training I had thought very seriously about going into paediatrics. I started work as a GP in 1984 and settled into my last practice in Southgate in 1987 where I remained until retirement.

Photo: Dr Lakhani and his wife getting ready to fly!

When and how did you first get involved with the charity?

I came to work in North London as a GP in 1987 and one of the first things I realised was that there were a high number of patients in the area with thalassaemia. Against conventional wisdom, I was shocked to see the percentage of thalassaemia patients showing up from the Asian communities. This was something I hadn't come across, even in paediatrics. My first real contact was on the phone to the society to find out how to support a patient. I became naturally interested in this condition, just to satisfy my own understanding of the subject. I soon realised the Society's offices were in Tottenham and I was based in Southgate, so my involvement developed further.

What made you decide to take the role of President?

Through my involvement I could see patients were experiencing all kinds of problems, some of which I could help with directly, given my medical background. Others were very challenging and hadn't been addressed. The Society was working in very difficult circumstances with limited resources and staff. I felt there was a lot I could give and the Society was asking me to do more and more things. By the time my predecessor stepped down from the role of President, I had been a Medical Advisor to UKTS for a while.

The society asked me if I would like to take on the role of President but they didn't really give me a choice, so I had to! I stayed on as President for 10 years from 1990-2000.

Can you describe what the charity was like when you were President?

To describe the charity, one really has to look at the patients. Back then, they were young, mainly adolescents, especially from the Mediterranean community. The Asians were younger, mostly babies. Treatment was far from ideal. The clinicians were doing a fantastic job but let's face it, we were giving treatment to prolong life that was slowly poisoning patients. Treatment was not easy; a subcutaneous pump driven injection. Many of the adolescents were struggling to cope with the daily injections and were suffering psychologically. Some stopped taking the injections which was a slow road to suicide, and it was heart-breaking to see these youngsters go through this. There was a big clamour to get the oral drug going. My predecessor had started on the road to look for an oral chelator agent as a treatment. There were complex issues around these drugs relating to patents and we had a complete morass of conflicting interests. We had to take the brave decision to say if the funding for the oral chelator came from the UKTS and the hard-earned efforts of parents of patients, then the patent belonged to the UKTS. It would have been hard for anyone to sue a poor charity and luckily they didn't try!

What were your biggest challenges at that time and how did you overcome these?

You could see when you met patients with thalassaemia that their treatment had not been perfect enough and that was a major challenge. Understandably, the charity was very much centred around the treatment aspects of the disease and we knew that we had to start to make inroads into other areas. It was a balancing act.

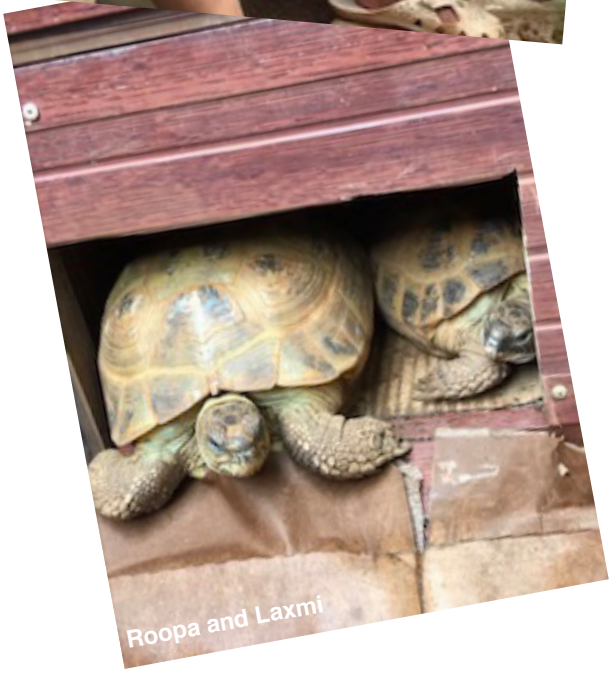
We were in Tottenham for a long time and we realised we needed a more high street profile. We were doing a lot to raise awareness at the time and by coincidence it happened that Southgate has a good mixture of all the affected communities. So we sought premises in Southgate and moved to where the Society's office is currently based.

What would you say were the main achievements during your time?

Moving the charity to the high street was very important in order to raise the profile of thalassaemia and educate people about the condition.

Awareness-raising and education. We wanted to make sure that all our publicity material was available the world over and were not interested in copyright. It was about the ethical use of medical knowledge for the patient.

We were doing serious work and looking for serious funding to start an Asian awareness



campaign here. We managed to get funding from the national lottery fund and undertook a three-year awareness campaign that was very successful. Following that, the rate of thalassaemic births, certainly in the South West and South of England dropped significantly.

When you look back, what makes you feel most proud?

The thing that really stands out for me is the confidence that I saw develop in the faces of patients. When I first came to the society, it appeared to me that there was a very high degree of anxiety and a fear of the future amongst patients and their parents. I think this is what touched me most initially. Over the span of the years, with all the progress we made, there was a metamorphosis from where we started to where we finished. In fact, I felt really proud that my successor was a patient with thalassaemia. The positivity had really changed.

Also, I believe I was also able to pull everybody together from the medical fraternity and I think the polarisation issues were not so major. On the national side of it, the fact that we came to be known as an international society because we were actively promoting education the world over was very good. And of course, the development of deferiprone.

How do you feel the charity has changed since then?

First and foremost, the ethos has remained. Our raison d'étre is the same: firstly, the welfare of patients; secondly, raising awareness; and thirdly, promoting research in the right areas. All of these things are still there and the foundations are very much intact. The other changes are cosmetic. We have lovely premises, more resource and are more evident to others in the street. I can see nothing but positivity and big pluses for the future!

We have a generation of patients with thalassaemia who have gone through adulthood

and now have confidence in their own health and in their future.

"I can see nothing but positivity and big pluses for the future!"

What do you think are the main issues we face now?

The biggest issue I think, if we step back from the UK, is the fact that we have so many thousands and thousands of patients the world over who don't yet have even the most basic treatments available. Obtaining a safe blood transfusion is the very first challenge. Saving your child from immediate death by blood transfusions is not a cheap thing for an average labourer in the third world. Chelation is even more expensive. If we look to work across the world, we have a mighty challenge but it is not impossible.

I used to think that our aim was to reduce birth rate down to zero as in Cyprus, but over the years I have realised the zero number is not important. What is important is that for any child born with thalassaemia, the parents have the fullest knowledge of the condition. I am very optimistic for the future because with the ability to genome study each child, more appropriate treatments will be in the offing.

What advice would you give to the UKTS team?

It is very important that you are seen to be approachable because you will have people with a variety of backgrounds and a variety of educational levels. Every person has an important story to tell. My other advice is to listen very, very carefully to those affected; parents and children.

If you know you have a point and are certain of what you hope to achieve, please do not give up, even if it means knocking on doors time and time again! I remember when we were trying to find patrons, we had many cases where we were turned down. If you have to knock on 100 doors and one of them opens, then you have come up trumps! You have to show the same resilience as the patients do, they are the ones who want to live and we have to make sure they can.

Similarly, what advice would you give people who have beta thalassaemia?

There is no better specialist for your condition than yourself. Never give up. As far as medical treatment is concerned, there will be more advances and you have a much better chance of living a full and healthy life into ripe old age.

What would you tell someone who is thinking about getting involved with the Society, perhaps by donating or volunteering?

Do it! Don't think twice about it!
The society has a small community it looks after but we have spawned a number of societies abroad. They have used our resources and literature and we want that, to educate the world because thalassaemia knows no borders, it is there the world over. So, my suggestion is yes, get involved!

This is a charity that has been value for money for all its past donors. It has been a charity that developed a drug that led to more and more research. And if you want numbers, just look at the age of the older patients! If you are looking to inspire yourself or others, come and join us!

"There is no better specialist for your condition than yourself. Never give up."

On a personal note, what are your interests and how do you like to spend your free time?

I retired four years ago and moved to Mauritius. Retirement has helped me to broaden my horizons. You might be surprised to hear that at the age of 61, I went and learnt to fly. It was a childhood dream of mine to be a pilot and I fly as often as I can – I have a simulator at home. I also play golf and I play the tabla (Indian drums) very badly! I like to travel too. I have three tortoises, my wife and I have had them for 25 years. I had tortoises as a child. As a Hindu, tortoises have a special significance in our religion. Krishna was a tortoise in a previous incarnation. When I got married, my mother-in-law's only marital advice to me was not to get a pet as it's passing would lead to my wife's endless crying! I took that very seriously and decided any pet would have to outlive us. The decision was made for us as a tortoise wandered in from next door and we had to look after it for a few days. That convinced my wife that we should get one and we ended up with three! They are originally from Kazakhstan so they love the tropical weather in Mauritius!

And who wouldn't love that tropical weather?! Thank you Dr Lakhani.

Thank you, farewell and good luck to Dr Anne Yardumian

As many of you will already know, the wonderful Dr Anne Yardumian retired in May this year. Whilst all of us at the UKTS were truly saddened to bid her farewell, we were delighted to honour her with a small reception, at the Head Office on 21 May 2019. It was a lovely evening attended by UKTS board and staff members, some of Dr Yardumian's thalassaemia patients and their families and some of her medical colleagues. There was a lot of reminiscing and fond old stories being recounted and we presented her with a keepsake book with photos from over the years and messages from all who knew her. Let's take a look at the amazing contribution Dr Yardumian has made to the world of thalassaemia over the years.

Until her recent retirement, Dr Yardumian had worked as a consultant haematologist at North Middlesex Hospital. She started working there on 1 January 1990. Earlier in her career, as a registrar she was seconded to Whittington Hospital as a medical registrar then went back there for a short time as a haematology registrar. It was whilst she was there in 1988-89 that she worked with Dr Wonke and took part in some of her red cell clinics. In Dr Yardumian's own words: "That's where I got the thalassaemia bug. It was Dr Wonke's clinics that really hooked my interest." When a job came up at North Middlesex Hospital shortly after that secondment, urged on by Dr Wonke, Dr Yardumian applied for the post and was appointed.

She notes that there were a number of sickle cell and thalassaemia patients when she started 29 years ago and she told us that: "One of the great delights in my continuing professional life is to see our lovely, now grown up-patients with thalassaemia and sickle cell whom I remember as children from 29 years ago, now with their own children!"

Dr Yardumian has also actually been involved with UKTS since 1990. As an advisor to UKTS, Dr Yardumian's work has been instrumental in creating pathways which objectively acknowledge the issues faced by patients living with thalassaemia, where these had previously been overlooked. Time and time again we have witnessed Dr Yardumian's inspirational drive to improve not just treatment for thalassaemia patients, but also strive to improve patients' quality of life.

We are incredibly grateful for her collaboration with us in creating the first UK and internationally recognised standards of clinical care of children and adults with thalassaemia in the UK in 2005. Not only was Dr Yardumian the Chair of the writing group for the first edition, she also gave herself the difficult task of chairing the next two editions. The standards manual was carefully written and compiled to provide easy access to specialist information on thalassaemia. Not only did the standards book act as a manual for medical professionals treating patients with thalassaemia, it became a tool which encouraged patients and

“One of the great delights in my continuing professional life is to see our lovely, now grown up-patients with thalassaemia and sickle cell whom I remember as children from 29 years ago, now with their own children!”

their loved ones to advocate for themselves to ensure they received proper treatment. We are hoping that we can 'borrow' Dr Yardumian from her retirement to help with the upcoming 4th version!

Dr Yardumian also invested a significant amount of her time in setting up the UK Forum and Peer Reviews for Haemoglobinopathies to try to provide support and correct the inequalities and lack of knowledge about thalassaemia observed across the country. As Dr Yardumian herself says: "The peer reviews have been pretty helpful in driving up the standards of care for people with thalassaemia and sickle cell across the country."

There was an overriding message from all the patients we spoke to; through Dr Yardumian's compassionate heart, she put smiles on many faces, gave patients the care and reassurance they needed to carry on and thrive in all aspects of their lives with thalassaemia. Many medical professionals also told us about how much of an inspiration Dr Yardumian has been to them throughout their careers.

Photo: Dr Yardumian, Dr Trompeter, Dr Kaya and Dr Drašar

Photo: Katerina Loizi (UKTS), Dr Yardumian and Maria Read

Photo: Dr Yardumian with Michael Michael (a former UKTS President) and his wife Aggie

Photo: Roanna Maharaj ((patient advocate) and Dr Inusa

Photo: Gina Halwani and Oddy Cooper (UKTS Treasurer)

On behalf of the UKTS and all of our members, thank you again to Dr Yardumian for her tremendous contribution to thalassaemia throughout her career. Not only has she been an excellent doctor, but she has also been an inspiring leader, mentor and a dear friend to many of us. Whilst we will miss her terribly, we wish her good luck and absolute success in all her future endeavours. Happy retirement!

GENE THERAPY...

These are exciting times in the world of thalassaemia research and development, with the recent European conditional approval for gene therapy potentially opening up new avenues of treatment. We are all eagerly waiting to see what changes these may bring, but with so many medical buzzwords flying around, it can get confusing! That's why we thought it would be a good time to look at the science behind gene therapy and explain the concepts in basic terms, so we can all understand what it means. We will start off by looking at what a gene actually is and then describe the process of gene therapy. This is a very simple overview and if you want to find out more, you can speak to your haematologist and get more information.

Please note that we are certainly not advocating the use of gene therapy; that is for you to decide (if you are eligible), under the guidance of your medical and nursing team. As with any new treatment, there are risks, challenges and unknowns but that discussion is not the purpose of this article here.

What is a gene?

Let's go back to the very basics and start by describing some biological concepts. Each of your cells has a nucleus that contain chromosomes. Chromosomes carry the genes that contain the instructions for making new cells in all the tissues and organs in your body, and then directing them to do their work.

A gene is a small packet of information that controls a characteristic in your body, by making very specialised protein molecules. It is a section of DNA, the unique molecule that makes up your chromosomes. Genes make us who we are; they determine our characteristics, for example, the colour of our hair, our height and so on.

..EXPLAINED

What does gene therapy aim to do?

In some cases a gene can be missing, defective or altered from the usual form. This can cause disease. In the case of a person with transfusion-dependent thalassaemia, they have inherited two altered ß-globin genes from their parents (one from the mother and one from the father). As a result of the altered – sometimes termed 'mutated' genes, the person cannot produce enough ß-globin, which forms part of haemoglobin. This is the substance in the red blood cell that carries oxygen.

How does gene therapy work?

There are three main approaches to gene therapy that are currently being studied:

- 'Switching off' the genes that are causing the problems.
- Removing the faulty genes and replacing them with functional ones.
- Adding functional genes to work correctly, alongside the faulty genes.

In the case of thalassaemia, it is the last method of gene addition that is currently being explored. The aim is to add copies of a functional gene so that haemoglobin production occurs effectively and the red blood cells are able to functional normally. A functional gene can be added inside (in vivo) or outside (ex vivo) the body. A patient can have an allogenic bone marrow transplant (BMT), where cells from another person, a donor, are used. Or the patient can have an autologous transplantation where their own cells are collected,treated and then returned to them. No donor is required.

Here we look at autologous transplantation BMT using the ex vivo scenario of gene therapy (the process is described on the next page).

Gene therapy: the process

Stage 1: Stem cell collection

The first stage of the gene therapy process is the collection of **stem cells**. What exactly are stem cells? Stem cells are blood cells at the earliest stage of development. All of our blood cells develop from stem cells in the bone marrow. Bone marrow is found inside our bones, mainly in the hip bone, spine, ribs and breast bone. Stem cells stay inside the bone marrow, mature and multiply and, when they are fully developed, they go into the bloodstream. Blood cells have a short life cycle. Every day the bone marrow normally makes millions of new blood cells to replace blood cells as they are needed. Stem cells are used because they have the ability to form many other types of cells in the body. You can think of them a bit like a blank canvas.

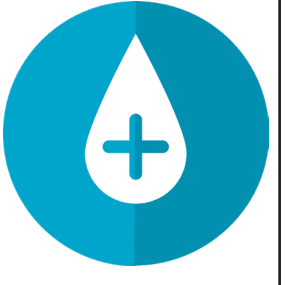
The first part in the process of collecting stem cells is to make them move from the bone marrow into the blood. This is called mobilisation (of the stem cells). Daily injections of a drug called a growth factor, stimulate the bone marrow and increase the number of stem cells it makes and help release them into the blood.

Apheresis is the name of the process to collect the stem cells. To do the actual physical collection of the stem cells, a cannula (thin tube) is inserted into a vein in each arm. Each cannula is connected by tubing to a machine called a cell separator. The blood travels through this sophisticated piece of machinery and is spun as it moves along. As it spins, the stem cells are separated out and collected in a bag. The rest of the blood and blood cells are returned to the patient through the cannula in the other arm.

Stage 2: Functional gene addition

This part of the process happens in a specialised laboratory. Functional copies of a healthy ß-globin gene is transferred into the harvested stem cells using a **vector** (which is effectively a vehicle). Often a virus is used as a vector but it has been genetically modified so that it does not cause infection. The 'corrected stem cells' or 'gene-modified cells' are then ready for infusion (insertion) into the patient.

Stage 3: Preparation treatment



The patient is prepared to receive the corrected stem cells through a process called **myeloablation** or myeloablative conditioning. This process uses high dose **chemotherapy** to wipe out the remaining bone marrow cells, to make way for those treated with the gene therapy.

Stage 4: Infusion and engraftment

Once the patient is ready after chemotherapy, the corrected stem cells are delivered by intravenous (IV, into the vein) infusion. After the gene therapy has been infused, the treated stem cells will need time to multiply. This process is called **engraftment**. In order for the production of high levels of haemoglobin, in other words, for the gene therapy to be successful, a significant number of genetically corrected stem cells need to be engrafted. Until they do settle in and multiply, the person's blood counts are very low and they are at risk of infection and may need antibiotics and/or blood transfusions, until recovery.

Stage 5: Recovery

The patient will remain in hospital until they have a sufficient number of new cells, their vital immune cells have returned to safe levels and their medical team deems it is time for them to be discharged. After the patient has recovered and is back home, they will continue to receive follow-up monitoring.

After the whole process is complete and if the gene therapy has worked successfully, the patient will be able to produce sufficient levels of haemoglobin. The process of gene therapy only needs to be administered once and if successful, this would mean that the patient no longer needs blood transfusions or chelation treatment.

It is important to note that some patients will experience a transfusion reduction, based on current data across all gene therapy programmes, so may benefit but not to the extent that they achieve freedom from transfusions.

All medical icons by Julie McMurry for Pixabay

Clearly, the outlook is promising and ongoing studies continue to identify the safest and best regimen for gene therapy treatment of thalassaemias. Watch this space!

Navigating periods of transitions

by Dr Chinea Eziefula

"Everything flows and nothing stays" – Heraclitus

The ancient Greek phrase 'panta rhei' coined by philosopher Heraclitus is roughly translated to mean everything flows. The message is that nothing stays the same and that change or transitions in life are inevitable. In this article, I am keen to focus on times of transition when living with thalassemia, because transitions can throw up a whole range of emotional reactions and behaviours; some helpful and others less so.

Autumn, in particular September, is a time of transition in many ways. For some it signifies changes in academic school years, with increased expectations when it comes to exams or homework assignments. It can highlight a significant move from primary to secondary school or a life change like going to university, starting a new job or searching for a path in the 'adult' world. Transitions can occur in many different aspects of life, at any point, and they can feel novel, exciting and thrilling!

Photo credit: Titouan Colomb on Unsplash

Interestingly, the physical sensations associated with excitement like a racing heart, tingling sensations in the body, feeling a warm buzz, getting a dry mouth or feeling light-headed (or 'dizzy with excitement') are also very similar to those physical sensations associated with anxiety and fear – the key difference between these sensations is simply how we understand and interpret these symptoms differently, depending on the context and the thoughts/ beliefs we have about them. Yes, going through transitions and big life changes can be exciting! However, such changes can also trigger a mixture of confusing and unpleasant emotions or, in some cases, an absence of feelings where people become 'numb' as a way of coping with the confusion and discomfort of change.

The common 'unpleasant' reactions to transitions and big life

Physical reactions	• ***Panic attacks*** • ***Unexplained chest pains*** • ***Headaches*** • ***Dizziness •Nightmares/ trouble sleeping due to troubling thoughts*** • ***Nausea*** • ***Constipation, diarrhoea or unusual changes in bowel movements*** • ***Fainting*** • ***Unusual aches/ pains/muscle tension*** • ***Frequent illness***
Emotional reactions	• ***Intense and persistent worries and fears (eg, not being able to "switch off" from thoughts about the future and what is to come, about past choices, about social pressures and the demand to meet expectations from others or from ourselves).*** • ***Irritability and feeling easily frustrated.*** • ***Unusual and frequent feelings of low mood and sadness.*** • ***Not feeling interested or excited about activities or interests that would usually excite and interest.***
Behavioural reactions	•***Trouble with memory and/or concentrating*** •***Over/under-eating*** •***Difficulties with decision-making*** •***Not taking care of appearance*** •***Not doing usual activities or interests*** •***Increased loneliness/ isolation*** •***Unusual smoking, drinking or other substance use*** •***Frequent tearfulness and crying*** •***Increased avoidance or procrastination*** •***Increased restlessness*** •***Frequent arguments***

There are various triggers for these confusing and unpleasant emotions. For instance, the pressures and demands that can occur during times of transition can affect people in different ways. Whilst we all have demands on us all of the time, it is when these demands and pressures become overwhelming that problems can arise. Academic stress or worry about performance at school or in work is very common with some reports of between 6-10 million schoolchildren experiencing this every year. Similarly, as many as 440,000 people in the UK report complaints of work-related stress, depression or anxiety that makes them ill (Heath and Safety Executive Statistics, 2015). Social comparisons can also be a source of stress or worry, particularly where an individual might worry about being perceived as different or 'less able' than their friends, peers or colleagues as is common when people have physical health and/or emotional health challenges.

If you have a long-term physical health condition like thalassaemia, there can be additional challenges that come with transitions into new environments (whether this is a new place of employment or study). These may include making decisions about whether or not to disclose, talk about or get support with studying or working.

Various styles of coping can result from these triggers and emotions. These include behaviours like pushing oneself so hard that self-care and self-management of health conditions like thalassaemia fall by the wayside. Other behaviours include extreme avoidance of situations that can be anxiety-provoking or patterns of ruminative thinking where people go over and over worries or thoughts about upsetting situations in their minds.

If you are going through these changes or you are noticing your child, friend or relative is becoming very anxious, low, withdrawn or they are worrying a lot about changes they might be going through, then ask them if they would like help and support with this. Consider the following tips and options to help or signpost you (or the person in need) to get help with understanding and navigating through any confusing or unpleasant feelings.

Support options

These support options may also help to overcome any fears, worries or other emotional challenges:

Remember 'panta rhei', everything flows and **change is inevitable.** It can be a good change or a bad change and how you cope with the change is the most important thing.

Try dealing with one thing at a time. Trying to tackle too many changes or demands at any one time is never a good idea. **Try to schedule and plan your time and activities** to allow you to cope better with each individual change or challenge. It's like when you are at school and you are asked to write an essay, you're often encouraged to start with an essay plan to help you think about where to start and to help you structure your essay well and complete it in good time. Do the same where you can with life's challenges and any big changes you are going through. Try to focus on one change at a time.

Wherever possible, **give yourself some time and space away** from pressures and demands – don't avoid them completely but have a healthy break away from them. This could be as little as a break of 5-minutes doing something fun, a calming or relaxing activity or something that distracts away from pressures and demands. You may need help to have a break so if there are people around you who can help you to do this then ask them to help you have a break! Here is a resource about living life with a long-term condition that is available via the NHS Moodzone website: https://llttf.com/home/long-term-conditions/reclaim-your-life/

Photo credit: Jill Wellington on Pixabay

Consider using some of the following online resources to help manage any troubling or unpleasant thoughts and feelings. There are many mobile phone applications and websites that focus on helping you to stay in the present moment and avoid getting caught up in unpleasant thoughts like Headspace https://www.headspace.com/how-it-works or Calm https://www.calm.com/.
There are also online therapy and support services for people who find it too hard to fit going to face-to-face appointments into their time, such as ieso digital online therapies https://www.iesohealth.com/en-gb or online community forums where you can share your thoughts and feelings with others who might be feeling the same like Big White Wall https://www.bigwhitewall.com/V2/About.aspx.

Please note that some of these resources may only be available in certain locations and the mobile applications mentioned offer free trials, although some sections of these platforms require further subscriptions or payments for full access.

Regular exercise or physical activity can help with stress, worry or unpleasant feelings or thoughts. This does not have to mean going to the gym or working out in ways that are intense. Ask your GP or healthcare professional about what is a safe level of exercise for you. For most people, some form of regular physical activity done at home, outside or in a sports facility can help. There is evidence to suggest that physical activity can release chemicals in your brain that tap into positive feelings such as happiness and can help people to feel energised.

See this NHS wellbeing guide for more information about the benefits of exercise and how to get started: https://www.nhs.uk/conditions/stress-anxiety-depression/mental-benefits-of-exercise/

Talk to schools, colleges or universities about 'pastoral support' or student counselling services. Ask employers if they have an employee assistance programmes (EAP). For specific advice and support about disclosing your condition consider talking to:

- Remploy: https://www.remploy.co.uk/
- Employment support programmes via Department of Work and Pensions job centres or via improving access to psychological therapies (IAPT) services (see below for more about how to access IAPT)
- Citizen's Advice Bureau: https://www.citizensadvice.org.uk/

Talk to a GP about any further forms of support.

Talk to your haematology team for help with managing life changes or transitions.

Ask for a referral to a haematology psychologist if this is available in your hospital.

Consider a self-referral to an NHS improving access to psychological therapies (IAPT) service: https://beta.nhs.uk/find-a-psychological-therapies-service/

Consider private psychological therapy options (if it is financially viable for you). When searching, always look for therapists who have been accredited by a recognised therapy association or organisation. In the UK the following websites host accredited therapists:

- British association for behavioural and cognitive psychotherapies - https://www.babcp.com/Public/What-is-CBT.aspx
- British association for counselling and psychotherapy: https://www.bacp.co.uk/
- Association for family therapy https://www.aft.org.uk/consider/view/family-therapy.html?tzcheck=1

If you are not sure if a private therapist is accredited in the UK, you can search the national health and care professions council register here: https://www.hcpc-uk.org/

Contact the UKTS to ask about support options and advice.

Dr Chinea Eziefula is a Clinical Psychologist and the Haematology Psychology Service Lead at Whittington Hospital. She will be writing a regular piece for our quarterly newsletter, so if you have any particular psychology-related topics that you would like her to write about, please send your suggestions to neelam@ukts.org.

Birmingham Patient Support Group

On 26 May this year, the UKTS ran a patient support group at Birmingham Hospital. Some 20 local patients attended the event and the feedback from them was very positive. We hope to run more patient support groups next year and will provide details via this magazine, our website and our social media channels, once dates and venues have been set.

In addition to a team of UKTS staff and Board Members, there were two consultant haematologists, nurses from the haematology unit at the hospital and two representatives from Oscar Sandwell Charity.

We were privileged to have two truly dedicated, specialist doctors attend the event and make presentations. Dr Shivan Pancham is Consultant Haematologist at Birmingham City Hospital. Dr Emma Drašar is a Consultant Haematologist at the University College Hospital London and Whittington Hospital. She is also the North Central Haemoglobinopathy Network Lead. She travelled to Birmingham especially for this event.

After some brief introductions and a plan for the day from the UKTS team, Dr Drašar made a presentation entitled 'looking good and living long.' Dr Pancham followed with a presentation about the latest treatments for thalassaemia currently being trialled; namely, Luspatercept and gene therapy. Each of the doctors' talks finished with question and answer sessions. Next came lunch which gave everyone an ideal chance to absorb the valuable information from the morning and also to get to know each other.

The afternoon session was less structured and gave patients a chance to discuss the issues pertinent to them. Dr Pancham spoke about the expected changes in commissioning with regard to the creation of haemoglobinopathy coordinating centres and

the National Haemoglobinopathy Registry (NHR). She stressed why it is important and helpful for patients to join the National Haemoglobinopathy Registry and how registering improves the standard of care across the entire NHS. Everyone was assured that the register protects their privacy and were given a demonstration of how the process was done online.

Community outreach and the UKTS ambassador programme were discussed, with clear agreement on the need for education. People talked about their experiences of thalassaemia with GPs, schools and employment. The common view was that more people need to know about the condition.

The involvement of the UKTS in the work of the newborn screening programme for sickle cell and thalassaemia was discussed as well as the importance of being screened and the implications of having positive results.

Most attendees were aware of screening but did not realise that everyone could be screened. They shared some ideas on who would be helpful to approach in the community and took some information to share amongst their family/ social groups.

From the patient feedback, a useful element in the day was the role-play between Dr Pancham

and UKTS Vice Chair, Anand Ghauttara. It was a role-play with a twist – more of a role reversal! Dr Pancham adopted the role of a young patient who was not taking their medication properly and Anand assumed the position of the doctor. Whilst it was extremely entertaining – if Dr Pancham ever wants a change in career, Hollywood awaits – there was an important message; chelation is crucial. Some of the patients were surprised that Dr Pancham recognises when they are being 'creative' in recounting their medication regimes and it made them see things from another perspective.

We learnt a lot over the course of the day and we hope all the patients found the day useful. We would like to say a huge thank you to the nurses for their work and time to make the event possible, in particular, Elizabeth Green, Haematology Unit Manager and Lead Nurse and Rejoice Nhevera (Liaison Sister). Thank you too to our wonderful doctors, Dr Drašar and Dr Pancham, for taking time to attend and for their excellent presentations.

We have summarised some of the key points from Dr Drašar's talk below. We have not included Dr Pancham's talks on treatment because she wrote an article on Luspatercept for the last issue of the magazine and we have featured an article on gene therapy in this issue (see page 18).

If you are currently on a trial for either of these treatments and would be willing to share your experiences, please contact neelam@ukts.org.

Dr Drašar: Looking good and living long

Dr Drašar talked about reactive medicine versus responsive medicine. In the past it was reactive, in the sense that doctors addressed issues as they arose. She emphasised that doctors are thinking ahead more now and trying to prevent problems before they occur. As a result, treatment has become more responsive. Dr Drašar noted that the most serious issue for transfused patients is iron overload; for non-transfused, it is the effects of chronic anaemia. She acknowledged that: "As a condition, thalassaemia can have effects on the whole body," but went on to make the point: "There is no reason for any difference in life expectancy for a person with thalassaemia."

Dr Drašar made everyone laugh when she brought up how doctors always check ferritin levels. She said, "We [doctors] are obsessed with it!" She then asked the question: "Why don't we always take chelation?" She noted that it can be difficult and "sometimes you just want to get on with your life without medication." However, she stressed the importance of adhering to a rigid chelation regime and discussed in detail the various symptoms and types of organ damage that can be incurred if this doesn't happen. Some patients were surprised to learn that not chelating properly can lead to a slate grey skin colour. Dr Drašar tried to drum home the important message: "Medication doesn't work in the box. Take your meds!"

Dr Drašar also touched upon the sensitive issue of mental health and how the hidden pain endured by patients with thalassaemia can lead to mental anguish. She recognised some people perceive a negative stigma associated with mental health matters and advised: "Talking makes so much difference. Get help and don't think others aren't going through what you are going through."

However, patients described how it is not always easy to access mental health services and how there is a degree of 'postcode lottery' (ie, certain areas have more resources and so accessing support can be dependent on where you live). Elizabeth Green, Haematology Unit Manager and Lead Nurse at Birmingham Hospital, advised patients - who may be struggling to get the help they need - to speak to one of their nurses, as they might be able to put them in touch with another patient who may be experiencing similar issues. She said that this kind of peer-to-peer support can often be very effective for patients. (For a list of contact points for mental health matters, please look at Dr Chinea's article in our last issue of the magazine entitled 'Thoughts, feelings and thalassaemia').

The key theme throughout the day was the importance of always medicating fully and properly. As Dr Drašar said, this is to live a full and healthy life whilst also maintaining your optimal appearance. The other key message was to make sure you speak up and advocate for yourself. Dr Pancham said: "I can never truly understand what it is like to have thalassaemia. Please tell your physician what your day-to-day struggles are."

"Part of the reason I did haematology is because of my family of patients. It's a relationship of trust. Be informed and aware of your condition. No one knows your thalassaemia like you know your thalassaemia. You are part of the team too."

Dr Emma Drašar

Photo: Dr Pancham and Dr Drašar

"The real heroes and champions of this partnership are the patients who we (the medical professionals) are humbled by, inspired by and grateful to have met."

Dr Shivan Pancham

Infected Blood Inquiry Update:
An interview with Jennifer Cole, OBE

With the second round of witness hearings due to start in October, we interviewed Jennifer Cole, Secretary to the Infected Blood Inquiry, to get the latest news.

Please provide a brief summary of your professional background.

Thank you for taking an interest in the Inquiry. I've served as a diplomat in Islamabad, Washington, Tehran and at NATO.

How and why did you join the Inquiry team?

Well, the idea of public service is very important to me. When I learned about the use of infected blood and blood products, I wanted to help people get answers as to why it happened. I've been with the Inquiry team since we began our work.

What is your role as part of Inquiry team?

As the Secretary to the Inquiry I support Sir Brian (the Chair) in his work.

Please can you give us a brief status update on where we are in the Inquiry process?

The Inquiry started by hearing from people infected and affected. Next year we'll start hearing from clinical experts and decision-makers. In the background, millions of documents from archives are being reviewed by our legal team.

How many people gave evidence at the first set of witness hearings?

Over 130 people gave evidence during nine weeks of hearings.

What have been the main sentiments coming through from the first set of witness hearings?

People infected and affected have spoken really powerfully about delays in being informed about infections, the stigma they've experienced, the poor communication by some healthcare professionals, difficulties in getting treatment, not being offered psychological support, and problems with financial support. There's been a real sense of solidarity supporting people giving evidence from everyone coming to the Inquiry and watching the hearings online.

What are the common themes being presented from the expert evidence currently in progress?

When we hear from clinical experts they will set out the current clinical knowledge about the viruses, blood and bleeding disorders and best practice in psychosocial support.

"Every person who speaks to the Inquiry about their own experience or their loved one's experience helps us to paint the complete picture of what happened."

Photo: Jennifer Cole

After the expert evidence, we understand there will be a second round of witness hearings. When do you expect these to be completed?

We will hear from decision-makers, including doctors, civil servants and politicians, which will take a number of months.

What happens after the second round of hearings?

People are at the heart of the Inquiry so just as we started with evidence from people infected and affected we are going to end with that too.

When does the Inquiry hope to conclude and what outcome do you anticipate at this stage?

The Inquiry report will set out what happened and why so that people infected and affected, as well as the wider public, finally have answers to their questions. Sir Brian will also make recommendations for the future. We want to be as quick as we can but we also know that people want us to be thorough in our work and not miss anything important.

We have members of our charity – people with thalassaemia who have been affected by the contaminated blood scandal – but who are hesitant about coming forward to present evidence. For some it is quite daunting and for others they do not think it will make any difference. What would be your advice to these members that are undecided about coming forward to present evidence?

Every person who speaks to the Inquiry about their own experience or their loved one's experience helps us to paint the complete picture of what happened.

There are several ways to do this. One way is to give a witness statement (which can be anonymous) with the support of the Inquiry team or your own solicitor. For people who don't want to give a statement, we have recruited trained professionals to act as 'intermediaries'. They visit people who request their support and then submit reports to the Inquiry covering a number of people's experiences, without revealing their identities.

The Inquiry team is available to help people decide what's right for them (Freephone 0808 169 1377 or contact@infectedbloodinquiry.org.uk). There's also a confidential support service run by a team from the British Red Cross, who have been working with the Inquiry since September 2018 (0800 458 9473 or 0203 417 0280).

Every witness statement or conversation with an intermediary is important. Each helps to paint the complete picture, including by supporting or confirming what others are saying. Sir Brian reads each and every statement and report. He is grateful to everyone giving a statement, speaking to the Inquiry's intermediaries, and supporting family members and friends who are participating in the Inquiry.

As a core participant, what can we at the UKTS do to support the work of the Inquiry?

As a core participant, you can help the Inquiry answer questions that people with thalassaemia have about what happened. You can also help by letting your members know about the opportunities to give evidence to the Inquiry.

Thank you to Jennifer and the Inquiry Team for a really useful update.

As Jennifer says, if you or a family member has been affected by this issue, we urge you to come forward and contact the Inquiry Team on the contacts listed above. Or please get in touch with UKTS and we can help.

Zaid Minhas: standing up to stigma

Meet Zaid Minhas, a young man who grew up in a culture where it was taboo to tell anyone you had thalassaemia or any other medical condition. If his family had relatives over to visit, he would have to remove his Desferrioxamine pump so his younger cousins wouldn't see or find out about his condition. Concealment took priority over treatment. Sadly, in some cultures, this stigma still exists today but we can draw inspiration from Zaid because, in his own words, "now my health comes first. Everyone and everything else comes second." Here is his story.

"I was born on 3 December 1988 in Lahore, Pakistan, the youngest of four siblings. Given that my parents hadn't heard of thalassaemia before, it came as a shock to them to receive the diagnosis when I was six months old that I had the condition. And so began my treatment regime at the Fatmid Foundation, a local charity-run unit for people with haemophilia and thalassaemia. It wasn't that straightforward though, because I was living in a society where you had to hide your condition; people were not very accepting. I lived in a joint family system at home where everyone else was 'normal'. Back then I remember I felt as if I had to blend in. But hiding it affects you mentally. I have come to terms with the fact that by hiding it you are making it worse for yourself.

When I was three, my aunt from England gifted me a pump to administer Desferrioxamine (an iron chelation medicine) – back then they were quite big units so they were not easy to hide. If we ever had to go out to visit relatives, I had to take the pump off beforehand, so in essence, that day I wouldn't have my treatment. If a relative was coming to visit, I would take the pump off before going to greet them. I was blessed that my parents could afford the medication in moderation. However, it saddens me to say that Pakistan, at least back then – was rampant with corruption. The long arm of this corrupt system extended even to the health industry and there were copious cases of fatalities caused by 'fake' medicines in circulation, so we were very mindful of that as well.

Photo: Zaid Minhas

The culture of 'no one needs to know' extended to school so you will not be surprised to hear, that none of my teachers or friends at primary or secondary school knew about my condition. I would attach my pump in the evening and wear it through the night and then take it off before school the next day so I wouldn't feel like an outcast if I was seen with it in school. In all honesty, I should have been wearing the pump every night but I

didn't like to self-inject so I tended to do it every other night instead. Furthermore, the blood transfusion process in Pakistan was never straightforward. Due to severe blood shortages, about 90% of the time you had to find your own blood donors. In my family, the only person who was a match for me was my dad. But he could not donate very often due to his age and also because he was then diagnosed with diabetes. This frustrated the whole process for me even more. To get around this, every two weeks my immediate family members would donate their blood to the hospital in exchange for blood for me. It was a system of 'blood for blood'.

My life changed completely when I came to visit my maternal aunt in the UK in 2011. My cousin suggested that I should consider studying in the UK and so I looked into the university system (UCAS) and did some research. I was given the opportunity to do an undergraduate degree at Anglia Ruskin University which I embraced with open arms. I rushed back to Lahore for just a week to organise my visa and to pack my bags! I completed the IELTS language course as it was a prerequisite to starting my undergraduate programme at Anglia Ruskin University, London.

After relocating to the UK to further pursue my education, what struck me immediately was how different the treatment in the UK was as compared to Pakistan. I was seen at Kings College Hospital (where I am still a patient) and it was strange to turn up to the hospital for a transfusion and to just be given blood! It felt so odd not having to bring along a blood donation from any of my family members. It felt so easy! Treatment at Kings has always been second to none.

It was strange being away from my family in a new country where everything was significantly different, but the level of treatment was so much better. I had a close circle of friends at university who knew about my condition. A year and a half into my BA (Bachelor programme) I started working at KFC as a cashier. After two and a half years, I was fitted with a PICC line at Kings. This was so much easier than having to self-inject the iron chelation drug, and that too without using any needles. But sadly, this was seen as a problem by my employer and I chose to resign.

Photo: Zaid at his graduation ceremony

After successfully graduating with an Upper Second Class in my Bachelor's degree from Anglia Ruskin University in 2014, I decided to study a Master's degree in International Project Management at the same institution. During this period I was living with my aunt. My elder sister and I have always been close and were living in the same house with our aunt until my sister got married in July 2015. My sister then moved out to live with her husband in Sutton, and I joined them. I experienced various hardships but I could cope because I had my sister with me. We were very close and she was like a mother to me. We supported each other mentally.

In August 2016, life took a shocking turn when my sister fell ill and was admitted to St. Helier Hospital. The doctors could not find the reason for her sudden sickness and admitted her into the hospital. Her condition deteriorated rapidly as days passed by. I still remember how difficult it was for me to talk to my parents and explain to them the situation at hand. When my parents realised the gravity of the situation, my mother caught an overnight flight to London to be with us. Unfortunately, on 10 August 2016, my sister passed away.

Devastated by the sudden death of my sister, this was an extremely tough period of my life. My family back home wanted answers too, as to what had led to her death. I had no answers to these questions, as the cause of my sister's death was yet to be determined by the coroner. I spoke to my medical team at Kings who were really helpful and gave me lots of options for support. I decided to get in touch with a charity called Uplift and attended group counselling sessions. The charity helped me a lot.

In hindsight, the easiest thing for me would have been to go back home to Pakistan – but then again, it would also have been the hardest. I say 'easiest' because I would have been with my family and I would have had their full help and support. On the other hand, I say 'hardest' because I was still awaiting the results of the coroner's report and I couldn't bear the thought of going home without the answer to these two questions. In addition, I didn't want to face the stigma again once and I would have had to rebuild my life once more. Shortly afterwards, I decided to move out from my sister's house as it was a constant reminder of her and didn't do me any good psychologically.

The coroner's report finally arrived in June 2017 and this was when we came to find out that the cause of death was cardiac failure. Those few years were extremely challenging for me as I undertook a lot of mental, emotional, psychological and financial stress. On top of this, I still had to overcome the challenges presented to me by my medical condition on a daily basis. However, the fact that I got through all of that and am still here today, made me realise how strong I really am and for the first time ever I appreciated my full potential. The world truly is your oyster, anything you set your mind to, you can achieve.

That brings me to where I am now, aged 30. I am in a better place mentally – I have a group of friends I am close with. I want to be active and get back to going to the gym. I am interested in film-making and enjoy editing videos on Final Cut Pro. I want to get a job and get on with my life. However, in stark contrast to my past, I no longer try to hide my thalassaemia or let anything else take priority over it. I have realised that health really is true wealth.

I am hoping to become a UKTS ambassador so I can help to educate people about what it's like living with this condition. I hope to inspire others by sharing my story with them. This has been the driving force behind my attempt to break the stigma surrounding such lifelong illnesses.Thank you for taking the time to hear my story."

Photo: Zaid with his late sister

Spotlight on Nepal

Our charity has strong links with the Nepal Thalassaemia Society (NTS), founded by Wendy Pinker and Durga Pathak (President). NTS was set up in the UK to raise funds for a blood transfusion centre in Nepal. The clinic now supports 140 children with thalassaemia - without NTS many of these children would have no hope for survival. The daily struggles these children and their families face to get basic treatment is heartbreaking and humbling to say the least. You can read their extraordinary stories, find out how the charity was created and how Wendy came to be involved at http://www.nepalts.org.uk/ourstory/. If you feel inspired by her incredible journey and the amazing work the charity is doing to help these children, please follow the link http://www.nepalts.org.uk/donation/ to donate. It is a truly worthy cause!

Photo: Children being transfused in Nepal

From left to right, (back row) Prem Man Byanjankar - senior technician Central Blood Bank, nurse Bagiswori Shrestha, Gabriel Theophanous, Wendy Pinker, Dr Manita Rajkarnikar - director Central Blood Bank, Dr Michael Angastiniotis, Durga Pathak (front row) Dr Sara Trompeter, Romaine Maharaj, patient Kamalnath UUpreti, Apo Pharma representative and Dr Prakash Yadav - Deputy Director Central Blood Bank.

Delegation visit

The Thalassaemia International Federation (TIF) and the UKTS jointly organised a delegation visit to Nepal, 4-6 August 2018. In addition to Wendy Pinker and Durga Pathak, the delegates included:

- Dr Michael Angastiniotis, Medical Advisor, Thalassaemia International Federation
- Dr Sara Trompeter, Consultant Haematologist and Paediatric Haematologist, Joint Red Cell Unit, University College London NHS Foundation Trust, Haematology Department, Cancer Services
- Gabriel Theophanous, UKTS President, TIF Board Member
- Romaine Maharaj, UKTS – Executive Director, TIF Board Member

There was also a representative from pharmaceutical company, Apo Pharma.

The purpose of the visit was to see the NTS clinic in operation and meet with the doctors and nurses who work there. However, the main aim was also to discuss with the Nepali government the case for better healthcare provision for thalassaemia patients in Nepal. It was a really productive trip with the local health authorities agreeing a short-term action plan:

- To provide free space in the state hospital for the treatment of children and adults.
- To make blood available to patients free of charge.
- To make chelation drugs free of charge to patients.

Long-term goals included:

- The agreement to conduct epidemiological studies in Nepal by the National Public Health Laboratory.
- The creation of a national thalassaemia registry to store thalassaemic patients' personal information and their medical history. This vital information will be a very important development for Nepal to help understand the disease better and thus also provide more effective treatment.

It's a year on since the visit so we thought it would be good to get a progress update from Wendy. This is what she told us: "The delegation visit had an enormous and positive impact on thalassaemia in Nepal. Free space is being negotiated. It may be time some time for this to happen but due to the delegation presenting a strong case, this is now on the government agenda. Since the visit, the government has pledged to make a budget for thalassaemia in Nepal. This is nothing short of a miracle! The details are not yet available. The delegation visit put massive pressure on the government to recognise and pay for the care of thalassaemia patients. NTS are hopeful that within the next five years, all diagnosed thalassaemia patients will be entitled to free blood transfusions in government hospitals. Free medicine may take a little longer. Prevalence will in turn be recorded."

Nurses' visit to the UK

One of the outcomes of the delegation visit, was the decision to bring nurses from Nepal to be trained in the UK.

In July this year, Bagiswori Shrestha and Yashoda Mahat, two dedicated thalassaemia nurses from the NTS clinic in Nepal came to the UK. Bagiswori has worked at the clinic for nine years and Yashoda for three years.

Under the guidance of Dr Sara Trompeter and her team, the nurses received training in the Haematology Department at University College London Hospital. They were also taught valuable skills from Clinical Nurse Specialist, Emma Prescott and her team at the Whittington Hospital. Some time was spent in the UKTS office learning more about the condition and Romaine Maharaj hosted them at weekends for sight-seeing trips.

According to Wendy, both nurses were overwhelmed by their first visit to the UK. Their training proved to be beyond their wildest dreams. They both felt that what they had learned was life-changing for them both and for the patients under their care. They are now the first ever UK trained thalassaemia nurses in Nepal.

Photo: Nurses Bagiswori Shrestha (second left) and Yashoda Mahat (in front) with nursing team and patient at the Whittington Hospital

Outlook

Whilst there is a long way to go, positive steps are being taken. Wendy is optimistic about the future and says: "There is great hope now that within a few years every diagnosed patient will have access to a full treatment of thalassaemia Both of our nurses have taken back their knowledge and will be a great asset to current and new thalassaemia patients alike. None of this would have been possible without the tremendous support of the UKTS, TIF, Sara Trompeter and Emma Prescott. We are forever indebted to them all."

Screen you genes on NATIONAL THALASSAEMIA DAY!

On Saturday 19 October, the UKTS will celebrate the first ever national thalassaemia Day! We have an open day in the office and will be inviting the general public to walk in and be tested for thalassaemia and other conditions. The UKTS will:

***

Offer free screening to the general public for thalassaemia and sickle cell trait.

***

Offer counselling and guidance (for positive test results)

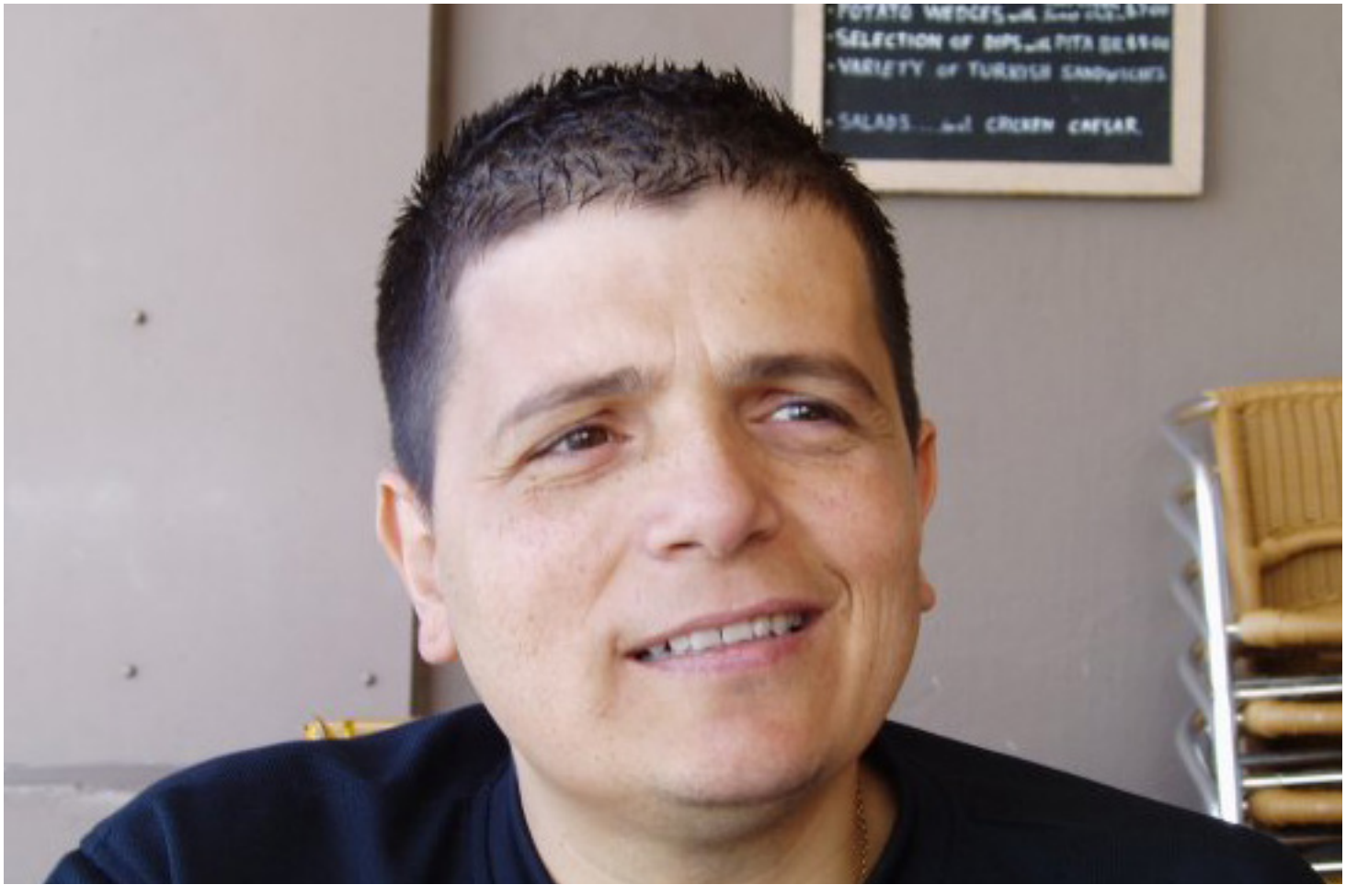
***

Partner with NHSBT for 'know your type' blood testing and to promote the need for blood donation.

Whilst these are the main activities the UKTS is concentrating on, additional events are being planned for the day. This is a major awareness-raising event. Given that there are over 300,000 carriers with thalassaemia in the United Kingdom and that it is a hereditary condition, clearly there are many members of the public who could benefit from thalassemia screening.

In memory of Costas Kountouras

We picked this particular date in honour of a very special person in the world of thalassaemia. It is the 10-year anniversary of the passing of Costas Kountourou. Costas was just a young man in his teens when he became one of the founding members of UKTS, way back in 1976. He was a pivotal figure at UKTS throughout his life from that point on. He was first elected to the UKTS Management Committee in 1982 as Assistant Public Relations Officer. Over the years, he went on to hold the positions of Secretary, Assistant Secretary and Vice President. He was greatly loved and valued by friends and colleagues alike for his kind heart, sense of fun and loyalty.



Below is a moving and heartfelt tribute by Gina Halwani, Costa's sister, that featured in the beautiful memorial book produced by the UKTS in November 2010.

A brother remembered

On 28th May 1961, a beautiful baby boy was born to Andreas and Mirianthi. He was named Costa after his maternal Grandfather. He was sadly diagnosed with thalassaemia at the age of only six months old and there his 48 year journey began. He lived in Camden with Mum, Dad and me, his 'big' sister. Growing up in Camden was great fun and there was never a dull moment with Costa around. The best part was all the friends he made and treasured. We often used to jokingly compare our friends and he would proudly say "there is no comparison, nothing like my friends, nothing like the boys". I wouldn't dare to disagree. He felt love and

respect for each and everyone of them. Too many to mention, his friends were also his family.

Another one of his loves in life was Arsenal. He followed games both home and away and insisted on his nephew Adam wearing his first baby grow bought from the Arsenal shop. He even swayed Mum to follow all the games on television and she used to prepare a comforting soup for him if Arsenal ever lost a game. He managed to attend school as often as he was able to and insisted on being treated the same as everyone else. His awareness of thalassaemia inspired him to begin fundraising for the cause and at the age of approximately 18 he joined the United Kingdom Thalassaemia Society and remained active on the Committee until his final days. He never ceased to encourage people everywhere to support the Annual Dinner and Dance which felt like his baby.

In between the dedicated work for the Society and his career in travel, he managed to find time to have the best possible social life that anyone could wish for. His 'little black book' was the best kept secret. I often used to get upset that he didn't manage to spend much time with me and the family, but now I understand. His life had to be shared with the many people and places that he had filled it with. The times I spent with him will be treasured in my heart forever.

At the age of 30, our father passed away and Costa took on the role of head of the family, a role he took seriously. Although he remained a confirmed bachelor, his nephews Adam and Louie, niece Roxan and Godsons were his children. Not forgetting all his friends' children that he became an uncle to. He lived his life to the full with vigour and passion and never allowed his illness to affect his ability to smile and keep on trekking like a real trooper. He was known as 'The Doc', 'TIPPA' and most fondly to me as 'Kotcho'. He was my strength when I was down and my friend when we laughed about the past. He was a man with strong morals and tradition and I always looked up to him. His word was the final word.

The void he leaves is unfathomable, but we must not forget his good work, his love for life, family and friends and we should endeavour to follow by example. If I could compare him to something, it would be to a multifaceted gem. Each side sparkling and as bright and wonderful as the other. He shone for each and every one of his friends and family, always assuring them of his friendship and love… We should remember him not how he died, but how he lived.
Forever in my heart and until we meet again...

Your sister Gina

We chatted to Gina about her brother and asked her what she thought of National Thalassaemia Day.

How do you feel about the date for National Thalassaemia Day being chosen to honour your brother?

I am overwhelmed with emotion, so very proud of his achievements throughout his short-lived time with us. I cannot put into words the feeling of knowing that his memory will live on in celebration of all that he did. Indeed, a wonderful way to honour and remember a great brother and friend.

What do you think about the activities planned for National Thalassaemia Day? What do you think Costas might have said about it?

Awareness is key and to be able to provide blood testing, screening and so on, to families that may not have the time or the ability to just walk in and get tested is of huge benefit. It is definitely a massive improvement on living with thalassaemia before. We have come a long way.

It is 10 years since the passing of your brother. Tell us how his memory has lived on for you and your family?

Costa lives with us every day of our lives, through his family and friends that he left behind. His legacy will live on because of the love, friendship and strong values he had. He instilled a sense of community not only amongst his Thalassaemia family but wherever he had the chance to meet people. His care and support stretched as far as the Middle East, Saudi Arabia, Asia, Australia and the list goes on. He felt very strongly about children and families not having the resources or the awareness to deal with this condition. He made lifelong friends along the way, some that have passed and some that are here today supporting me and my family. How could we ever forget - he was larger than life itself!

Is there anything else you would like to add?

I could write a book about Costa and my life as his sister - but a few choice words can cover the wonderful, kind and memorable person that he was. I will never be able to fill his shoes, but I will do my best to continue working with this thalassaemia family as best as I can. My family and I are truly honoured by this special tribute.

Meet Valdir, the running professor

We are very excited to report that our spaces for next year's London Marathon are filling up quickly! We will be featuring our runners in upcoming issues of the magazine but we wanted to start with this runner, Valdir de Andrade Braga, because he has already managed to raise over £800 for our charity! Find out why Valdir is keen to support us.

My name is Valdir de Andrade Braga and I am a professor and researcher at the Federal University of Paraibain Brazil, where I teach Human Physiology. My research is focused on understanding the mechanisms by which people develop high blood pressure, diabetes, obesity, and other metabolic diseases. Over the last 10 years, I have published over 100 research articles on these topics. The purpose of my research is to provide the scientific background to convince patients that a healthy lifestyle, including diet and exercise, is good for them.

In my own endeavour for a better life, I started running two years ago. Since then, I have run four marathons, all of them in under three hours. My ultimate goal is to complete the Abbott Marathon Major Series. For that, I have already completed Boston, Berlin and New York City.

Most of the sponsorship will be given by family and friends. I will also be helped by my running club, Fit Runners, who are very active in supporting humanitarian and charitable causes. Additionally, I will use my own funds and personal social media to gather potential sponsors.

The fact that thalassaemia is a blood disorder that affects more than 280 million people in the world caught my attention and I felt I wanted to contribute somehow. Therefore the opportunity to combine my competitive amateur athlete life with a cause that is related to my research, is a perfect match. I am thrilled to be representing the Society at the 2020 Virgin London Marathon.

Well done and thank you Valdir!

United Kingdom Thalassaemia Society

-STANDING ORDER MANDATE-

Help us to help you by supporting YOUR Society
-Every £1 is precious!

Please Support The UK Thalassaemia Society
by Making a Monthly Donation

To the Manager [Name of Your Bank]	
Address:	
City:	Postcode:

Please Pay: NatWest, 12 The Broadway, Southgate, London N14 6PL

For the credit of: UK Thalassaemia Society, Registered Charity No:275107
Sort Code: 51-50-00 Account Number: 64949362

The sum of: £2.00 ☐ £5.00 ☐ £10.00 ☐ Other ☐ £_____ (amount)

On the_____ (day) _____ (month) _____ (year)

And thereafter every month until further notice and debit my account accordingly.

Signed:	Date:
Signed:	Date:

I would like tax to be reclaimed on my donation under the **Gift Aid Scheme**. I am a UK tax payer and pay an amount of income tax and/or capital gains tax at least equal to the tax that can be reclaimed on my donation.

YES ☐ NO ☐

Data Protection

ALL DETAILS AND INFORMATION WILL BE KEPT ON OUR COMPUTERS AND WILL REMAIN IN THE OFFICE. YOUR INFORMATION WILL NOT BE MADE AVAILABLE TO ANYONE OUTSIDE OF THE UKTS.

I AGREE FOR MY PERSONAL DETAILS TO BE KEPT AT THE UKTS OFFICE AND ONLY BE ACCESSED BY UKTS OFFICIALS. I UNDERSTAND THAT I CAN HAVE MY PERSONAL INFORMATION REMOVED WHENEVER I WISH.

YES ☐ NO ☐

Please contact us if you have any queries. When completed, please return form to the UKTS office (see below). We will then send this form on to your bank.
Thank You for your valued support.

In memory of Loukia Kanias

We would like to extend our condolences to Koula Kanias and Christine Christodoulou on the passing of their mother, Loukia. She was an incredible, determined, selfless and caring woman who inspired every soul she met. We are honoured to have known her. We wish Christine, Koula and the whole family all the strength they need to get through this difficult time. Thank you to them all for raising over £800 for the UKTS in her name.

Photo: Loukia with Panayiotis

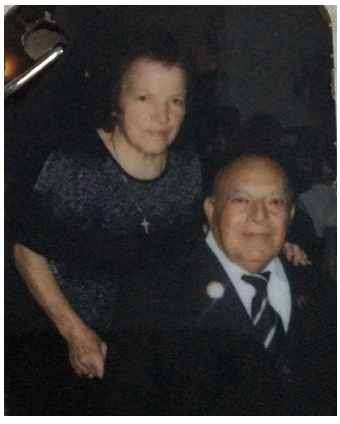


Photo: Loukia with daughters, Christine and Koula (as a young family in photo above)

Christine tells us about her parents.
"My father Panayiotis came to the UK from a village in Cyprus called Yalousa in 1949. He came here to pursue his dream for an easier and more prosperous life than Cyprus. He then returned to Cyprus in 1961 and was introduced to his wife to be; my mother Loukia, who was from Tavros. They both returned to the UK and got married in Camden Greek Orthodox Church on Prat Street, on 5 November 1961.

Their first child, Koula, was born on 1 August 1962, their second child, Frosa, on 22 July 1963 and their third child Maria on 6 April 1965. Then came identical twins Michael and Andrew born on 18 March 1966 and lastly me, Christine on 19 November 1970.

My sister Koula was diagnosed with thalassaemia major at the age of one and started her blood transfusions. My mum continued having kids, not totally understanding what thalassaemia meant. After having the twins, the doctor told her not to have any more children. So when she was pregnant with me, she used to go to the hospital for my sister Koula and hide her pregnancy.

So I was born and at the age of seven months I became well. My mum was too scared to speak to the doctor, but had no choice. The doctor asked to bring me in for testing and it was confirmed that I also had thalassaemia major. I started my blood transfusions at the age of two."

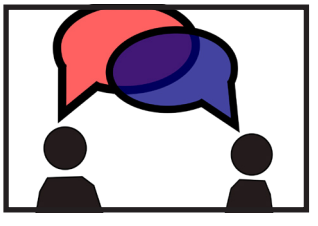
DATES FOR YOUR DIARY

There are two really BIG and exciting events on the calendar for the thalassaemia community before the end of 2019, so put them in your diaries now!

19 October 2019 - The first is our National Thalassaemia Day featured on p42 where, for the first time ever, we will be opening the office to the general public and inviting everyone to come in and be tested for thalassaemia and other conditions. Refreshments will be served.

7 December 2019 - The second is our annual fundraiser which should be a very merry way to kick off the festive period! Get your gladrags on and come ready to party the night away! Get your tickets before it's too late...

Please do join with us and support these events - it's a great way to meet other people in the thalassaemia community, raise much-needed awareness of the condition, support the charity whilst having fun!



Tell us what you think about this issue of Thalassaemia Matters. Email the editor Neelam Dongha on neelam@ukts.org

Would you like to share your story? Or do you have any article ideas that you would like to suggest for future issues of Thalassaemia Matters? We would love to hear from you. Please do email the editor with your comments and suggestions.


United Kingdom Thalassaemia Society
Invites you to a fun filled evening
Festive Charity Gala
7th December 2019 | 18:30 - 12:00
The Hive London, Camrose Avenue, London HA8 6AG
Drinks Reception
Three course dinner, drinks and entertainment
Auction hosted by Chris Tarrant OBE
Tombola & Treats | Dancers & DJ
Dress Code - Cocktail
Tickets - £100 per person
Tel: 020 8882 0011
CHARITY REG NO 275107
www.ukts.com

United Kingdom Thalassaemia Society

-Membership Sign Up-

Being a member gives you: Regular newsletters to keep you up to date on national developments, Invitation to our events, right to vote at our AGM to decide how the society is run, join the UKs largest community of people affected by thalassaemia

Personal Details

Title (Mr/Ms/Dr):

First Name(s):

Surname:

Address:

Postcode:

Occupation:

Ethnic Origin: (Optional)

Contact Details

Telephone (home):

Telephone (work):

Mobile:

Email:

Are you a (please tick where appropriate):

Patient: ☐ Parent: ☐ Organisation: ☐

Health Care Professional: ☐ Other (Please state): _____

If you are a patient or parent, please complete below:

Patient's Names:

Date of Birth:

Gender you identify with:

Type of thalassaemia (Major, intermedia etc):

Hospital where treated:

Important: We can only keep your details if you give us permission. Please tick yes if you would like to receive any further contact from us.

All details and information will be kept on our files. It will not be made available to anyone outside of the UKTS.

I agree for my personal details to be held by the UKTS and for the UKTS to use these details to contact me about their work and issues relating to thalassaemia.

Yes: ☐ **No:** ☐

UK Thalassaemia Society, 19 The Broadway, London N14 6PH
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