UNITED KINGDOM
Thalassaemia SOCIETY
matters
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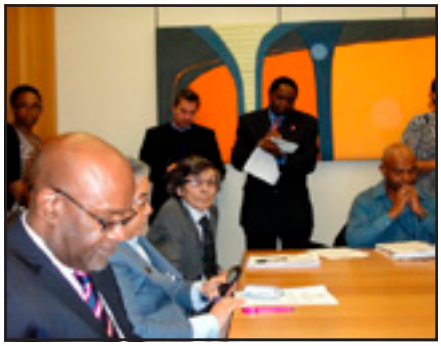
...continuing the fight against thalassaemia

All Party Parliamentary Group discusses inconsistencies in care



UKTS Secretary Mr George Constantinou (centre, looking at camera) at the APPG meeting. In the foreground is Mr John James, Chief Executive of the Sickle Cell Society

The All Party Parliamentary Group for Sickle cell and Thalassaemia met on the 25th November 2013 at Portcullis House, Westminster. The UK Thalassaemia Society was represented by UKTS Secretary George Constantinou and patient member Roanna Maharaj. The meeting focussed on the overview report from the recently completed peer review of adult services for thalassaemia and sickle cell, which was conducted by the UK Forum for Haemoglobin Disorders with the assistance of the West Midlands Quality Review Service. The Clinical Leads of the peer review, Dr Kate Ryan (Consultant Haematologist, Manchester Royal Infirmary) and Dr Jo Howard (Consultant Haematologist, Guy's & St Thomas's Hospitals) were present; as was the Chair of the UK Forum on Haemoglobin Disorders Dr Anne Yardumian (Consultant Haematologist, North Middlesex Hospital). The meeting was chaired by Diane Abbott MP; and was attended by a range of stakeholders including Parliamentarians from both houses, individuals with sickle cell and thalassaemia, carers, representatives from the voluntary sector and medical professionals.

The findings of the adult peer review were broadly similar to those of the review of paediatric services in 2010/11. The overview report made it clear that inconsistency is the defining feature of services for the haemoglobinopathies. The quality of the facilities alone is extremely variable, with a few services having extremely high quality premises and facilities, while others are unfit for purpose due to lack of space and/or basic equipment. In addition, there was very little evidence of forward planning, even where there are large numbers of children who will be expected to transition to adult services. Even though psychology and social care support are considered to be essential elements of thalassaemia and sickle cell care, the majority of specialist teams had no access to a specific psychologist or named social worker. Of particular relevance to thalassaemia patients is the fact that many patients have no access to treatment outside office hours and regularly miss one and a half days of education or employment a month, or have to take them as annual leave. Several APPG participants commented on the contrast with other services, for example cancer, which is now extremely well resourced. The haemoglobinopathies, unfortunately, appear to be ten years behind the cancer services, despite the fact that guidelines and standards for treatment have been in existence for many years. The overview report did however pay tribute to the extreme dedication and tireless work of the medical and nursing staff; and acknowledged that most services could not run at all if it they were not prepared to go above and beyond what is strictly in their job descriptions – a position that we in the thalassaemia community can wholeheartedly endorse.

It was agreed that Diane Abbott MP and David Burrowes MP (Chair and Vice-Chair of the APPG) will write to the Medical Director of NHS England pointing out the findings of the peer review reports and asking what action will be taken to improve and support thalassaemia and sickle cell services; including funding for future peer reviews, action to implement the recommendations of the reviews and better education and training in the haemoglobinopathies for health care professionals.

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A word from our President

It has been a busy time since the last issue – not only have we had the Thalassaemia International Federation conference in Abu Dhabi, but there has been a lot of the ongoing behind-the-scenes work of meetings and policy discussions which are an essential part of running a national charity.

First of all, though, let me say without hesitation what a marvellous time we had in Abu Dhabi – I think that any of the UK patients and parents who attended would agree that it was a unique experience, the venue was fantastic and the hospitality was second to none. Best of all however was having the chance to meet up with old friends from past international conferences and catching up with their news – and meeting new people, some of them parents with young children who have thalassaemia. As a thalassaemia patient myself, one of the most rewarding things about working with UKTS is meeting new parents, talking to them about my life and showing them that I am a regular guy – so that hopefully they will go away knowing that thalassaemia, with all its challenges, does not stop us from living a normal life.

On page 5 you will find a fascinating article by Gagandeep Khattar, who has put so much of his own time and effort into developing an electronic patient record specifically designed with thalassaemia in mind. This project created a great deal of interest in Abu Dhabi from patients and medical staff alike. Watch this space for further developments.

Another person I really enjoyed meeting in Abu Dhabi was Josephine Bila, a thalassaemia patient from the USA who gave an inspiring presentation – she has kindly agreed to put it into article format and you can read it for yourself on page 6 and while we're on the subject of inspiring patients, don't miss the article on page 10 by our own Roanna Maharaj, who is originally from the Caribbean but has now made her home in London. I think you will agree that their loss is our gain!

At the beginning of this article I mentioned the many meetings and discussions which are part of the work of the UKTS management committee. On the opposite page you will see the announcement of our next AGM; and I invite any UKTS member who wants to make their own contribution to thalassaemia in the UK to put their name forward for election to the committee. We welcome new ideas and we could really do with some fresh blood (sorry, I couldn't resist!). Seriously, though, if you feel you have something to say, why not give it a try? Just contact our staff members Elaine or Katie if you want any further information.

Wishing everyone well over the festive season and I hope to see you at the AGM!

With warmest good wishes,

Gabriel Theophanous
President, UK Thalassaemia Society

Our Mission Statement

To be the definitive source of information, education and research for those affected by, or working with thalassaemia.

The UKTS Management Committee

President Gabriel Theophanous	*Treasurer* Romaine Maharaj
Vice-President Mike Michael	*Assistant Treasurer* Pany Garibaldinos
Secretary George Constantinou	*Committee Member* Nina Demetriou
Assistant Secretary Andy Charalambous	

AGM Announcement

The UK Thalassaemia Society AGM will be held at 7.30pm 21st January 2014 at the UKTS office

19 The Broadway, Southgate, London N14 6PH

All UKTS members are cordially invited.

Many of you have emailed the UKTS office to say how much you enjoyed the film **"Building Blood"** which reported on the current research into the manufacture of red blood cells and how this development could benefit thalassaemia patients. We are delighted to announce that Professor Marc Turner, one of the lead researchers on the project, has kindly agreed to attend the AGM and give a presentation to our members. Don't miss this chance to get a first-hand update on this project, which could ultimately lead to fewer transfusions and therefore a reduction in iron overload.

Light refreshments will be served after the AGM. If you are planning to attend please notify the UKTS office to enable us to make catering arrangements.

If you are a UKTS member you are eligible to put your name forward for election to the UKTS management committee.

YOUR SOCIETY NEEDS YOU!

Join the UKTS management committee

No previous experience necessary – just enthusiasm and commitment!

Please see the enclosed letter and nomination form. Please note that all nominations must be with the UKTS office by 5th January 2014.

TIF 2013 Abu Dhabi

Thalassemia International
Federation World Congress

What goes on in Abu Dhabi stays in Abu Dhabi

By Chris Fassis

Chris (far right) et al at Viceroy Hotel Yas Marina

But this much I can tell you. For those of you that have never been to a TIF conference you have seriously missed out on making some really exceptional friends and some lasting relationships albeit distant ones.

Attending a conference is like an incredible emotional rollercoaster filled with many highs and sadly a few lows; it is the opportunity to see how your fellow thalassaemic friend copes under a variety of circumstances and how he or she balances life within these specific constraints.

However it's outside of the conference hall after you absorbed some of the talks, listened to various ideas and perhaps even momentarily nodded off that the real conference begins, especially at dusk around the dinner table or even more importantly at the bar!"

Yes you can drink. .....responsibly of course!! And you would not believe the subjects that one can cover in such a short time , anything from discussing the merits of bone marrow transplant to telling your friends how the plight of not receiving a well known chelator in another part of the world was remedied simply by putting one person in touch with another or about the two sets of new parents that found a quiet corner in the conference centre and spent several hours just chatting together and sharing dreams and aspirations for their little ones. THIS is the essence of conference, and all of this was / is being carried out against a backdrop of pure opulence, spectacular buildings and supercars.

However Abu Dhabi is more than glitzy buildings and flash cars it too has its fair share of thalassaemic patients and there lies our commonality in that thalassaemia can be found in a rich neighbourhood or a poor one and that the one thing that links us all is that we all hope for a better tomorrow and by pulling together we will achieve this.

At this point I would like to take the opportunity to thank everyone that helped to make TIF 2013 such a success from the attendees, the sponsors, the various societies from around the world and most importantly our host His Royal Highness Dr Sheikh Sultan Bin Khalifa A Bin Zayed Al Nahyan for making this all possible. I must say that was one wonderful gala dinner we had at your Palace. What a night! From the minute we arrived to the moment we left, unbelievable!! Such a choice of food and the cakes….. oh my the cakes they were incredibly yummy I was in the cake room so long I almost missed the bus back to the hotel ! I know I know I'm diabetic but hey you know it would be rude not to!

I hope that as you read this you get a sense that conference is fun, informative inspiring and thought provoking and above all else you should begin to see how your own efforts at home wherever home is; as you sit down to take your chelation or treatment that your efforts have a rippling effect on your fellow thalassaemic and that by your own endeavours you give others the incentive and realisation that they too can achieve their dreams of a long life, their own family and that any dream they may hold can be helped to become reality.

Thank you for taking the time to read my brief insight to why TIF conferences are important to attend.

Track Your Health – Journey of an App in Abu Dhabi

By Gagandeep Khattar

Gagan (foreground) at the conference; with UKTS President Gabriel Theophanous and Vice-President Mike Michael to his left.

Being a responsible human being and a thalassaemic, I have always wanted to give back what I have received from society. On one of my appointments with specialist consultant Dr. Shah, I showed my interest and desire to contribute something helpful; and she encouraged me to do whatever I could do best. The solution which came to my mind was to use my hands-on experience in IT, (information technology), a field which connects people, is easily approachable and just a click away. Software development is the skill which gives me my livelihood and this seemed to be the best way I could make a contribution for my fellow thalassaemics. I conceived a concept for an electronic health tracker application in March 2013 and started working on developing a prototype in May. The prototype is ready now and is called: **trackurhealth.co.uk**

Now let me tell you what the app is all about. This app will be a one stop shop, a website, iPhone and Android app, which will help every thalassaemic in making their life easier, giving them control about their health and encouraging them to lead a more confident life in society. This app will bridge the gap between thalassaemics from the developing and developed worlds. The app empowers each and every thalassaemic to use the power of IT to manage their tests, reports, range, transfusion day data, iron chelation, allergies etc. by just a click. By using the app, reminders and alerts can be set for medicines, appointments, vaccinations etc. Every patient will have the privilege of comparing their own result with the given range, baselined to the world's best hospitals like the Whittington or high standards set by world class patient societies like UKTS. Also patients will be able to use features to see news and updates, chat amongst themselves and write blogs. Once the app is fully launched, patients can log on and use the app from any device, i.e. laptop, iPhone, Android etc.

Security of data is of course a key concern. To provide the highest level of security, anonymity of patients will be maintained and encryption will be done on key data. Two levels of email verification will be in place and data will be kept within the UK on a dedicated host server in which there will be no shared firewall or sharing with other applications.

With the help of Mike Michael, Vice-President of UKTS, I had the opportunity to present my app to the TIF board in the presence of Her Highness Sheikha Sheikha Bint Seif Al-Nahyan. The idea was very well received by TIF Board and their Executive Director, Dr Androulla Eleftheriou, has invited me with UKTS to put forward a formal proposal for building the complete app. In Abu Dhabi, I discussed the app with many other participants from other countries, who were patients, parents and delegates. Maria Kastoras of Thalassaemia Australia along with many other participants promised me full support in promoting awareness for the app when it is ready for launch. Also Helen Ziavra, President of the Thalassaemia Foundation of Canada who is also a TIF board member invited me to present the app to her society in Canada. It was really a very nice experience to share the idea of the app with so many of my fellow thalassaemics in one place. I would like to take this opportunity to say thanks to UKTS members Gabriel Theophanous, Raj Klair, Romaine Maharaj, George Constantinou and Mike Michael who gave me loads of support and helped me to prepare for the presentation.

Believe in Yourself

Josephine Bila is a thalassaemia major patient from the USA who gave a presentation at the recent TIF conference in Abu Dhabi. She has very kindly converted her presentation into an article format so that it can be shared with our readers.

Josephine Bila speaking at the TIF conference October 2013

When I was a kid, my parents didn't want me to talk about thalassemia. They wanted me to separate my hospital life from my home life. I believe this was their way of protecting me. Unfortunately, hiding the truth of who I was really hurt my self-esteem. I felt terrible about myself and always worried what people would think if they ever found out.

At the age of seven, I lost my best friend at the hospital. She had haemophilia. That's when I became very aware of my own mortality. By the age of eight, I was going to the hospital every week for a transfusion. I was a shy and never talked about being sick. My schoolwork suffered due to my poor attention span. Because of this, I was placed in special needs classes for reading, math, and writing.

The winter after my 8th birthday, I got the flu and was put into the hospital for several days. On the fourth day of my stay, my doctor came in to talk to my mother. They appeared to be whispering at the foot of my bed. This immediately made me think that they were plotting out what to do if I became gravely ill from the flu. As my doctor was leaving the room, I asked him, "Am I going to die?" The doctor laughed and said, "Not any time soon, kid." Then he said, "Why don't you get out of this bed and go for a little walk? You can probably go home tomorrow."

Go home tomorrow? I'm not going to die? I couldn't believe it. My mom said, "Let's go. A nice lady down the hall wants to meet you. Her baby, Pai, lives in the hospital." WHAT! I was horrified to learn that a baby was LIVING in the hospital.

I really didn't want to see this baby or her mother, but I agreed to go because I felt bad. When I got to the room, the woman was very kind. She said "Pai is in the crib; go say hello." I walked over to

the crib, got on my tippy toes, grabbed the metal bars, and peeked over the top. The baby did the same. She grabbed onto the bars and looked at me. That's when I realized that Pai had a tube coming out of her chest and another going into her nose. I got so scared that I backed away from the crib.

Instead of getting scared of me, Pai lifted her arms up and gave me a really big baby laugh. Her laugh released me from my fear. That was a big lesson for me. Pai taught me that I should not be afraid of things to come. She gave me a new perspective on life and made me realize that I didn't have to constantly feel sorry for myself or be afraid of dying.

The next day, I was allowed to leave the hospital just as my doctor predicted. As the doors opened up to the outside world, I saw the sky for the first time in days. I took a deep breath in and I realized that I was experiencing something Pai may never again experience. I was free.

Even though I was released from some deep-rooted fears, I was still miserable. My teen years were the worst. None of my friends knew I was going to the doctor. I wasn't even sure if my sister knew I was getting transfusions. All of this together made me feel horrible about myself. Of course, I missed so much school that my teachers had to know why I was absent. Some of my teachers were really mean to me about it. My 6th grade teacher asked me how I could possibly smile so often yet have such a tragic life. Another teacher told me that I would never amount to anything. And in 11th grade, when it came time for my guidance counsellor to help me find a college, she said that no college would accept me, so I shouldn't bother trying.

Everyone's negativity hurt me. My grades suffered for a long time because I believed all of the bad things people were saying. By the time I reached 10th grade, I had a 67 out of 100 grade point average. My father never paid attention to my report cards. He just kept drilling into my brain that I needed to keep trying. He'd say "school is everything." My dad knew school was important because neither he, my mom, nor anyone in my extended family went to college. As my parents pushed me to succeed in school, I started to believe in myself and slowly stopped believing the mean things teachers and kids were saying about me. Their influence worked. I soon began to succeed.

What I learned is that you are the designer of your life. Other people will try to label you and make you feel like your life is worth less than theirs and that's absolutely untrue. You have to let go of other people's beliefs and create a bigger belief about yourself.

This leads me to the first of three big lessons that I want to share with you. First, I want you to talk about thalassemia. Align yourself with people who are compassionate and will understand and appreciate the things you're going through. These people could be family members, nurses at your hospital, or close friends. People who know and care about you.

I didn't start opening up to friends or anyone else until I was about 26 years old. Living in this type of isolation was so stressful that it probably took years off of my life. What I didn't realize was how incredibly freeing it is to actually stand in my truth and not hide who I am. When I finally opened up to my closest friends and asked my sister if she knew what was going on, I felt a great weight lift off my shoulders. I didn't have to hide anymore. Of course, not everyone reacted nicely, but it didn't matter. I gave myself permission to just be me. The more people I tell, the less I see thalassemia as my identity.

My second lesson: Don't take the easy way out and say you shouldn't achieve goals because you're just going to die anyway. Engage in activities that bring you alive and keep you intrigued. Since I am a science geek, I will use Stephen Hawking as a prime example here. Stephen Hawking is known to the world as a genius who studies the universe as a cosmologist. At the age of 21, his doctors diagnosed him with a motor neuron disease and told him that he only had two years to live. Do you know how old Stephen Hawking is today? He's 71! Do you know why he lived three years, four years, and now fifty years beyond his diagnosis? Because he lives with absolute passion. Mr. Hawking is in love with our universe and that love fills him with enough curiosity to keep him looking forward to each new day of life.

Lesson three: You will absolutely lose out on life if you listen to other people's negativity. One of my clients has thalassemia and said "Josephine, I wake up in the morning, look in the mirror and all I see is thalassemia." That was him deeply identifying with his illness. After working together, he realized how much he was allowing other people to negatively influence his perception of himself. He is now learning to embrace the things that make him unique. We are uncovering his passions and creating small achievements that will help him build momentum to create a new way of seeing himself.

I'm 37 and still learning how powerful I can be when I live in my truth. Living in my truth means telling people that I have thalassemia, listening to my inner desires about achieving goals, and not allowing anyone else's beliefs to impact my identity. Since I began to just be me, without the fears, worries, and negative belief system, I was able to graduate college and graduate school; I was hired by the United States Government to be the voice of thalassemia patients in America; I created a successful blog and wrote a book, all while working for one of the top entertainment companies in the world.

Your dreams belong to you for a reason. Put faith in yourself and not in the words of those who try to deny your life of true meaning. Love yourself first and everything good will follow.

Josephine Bila, M.S.W., is an international speaker and health coach. She works with people who have chronic illness, freeing them of their anxieties, fears and sadness. If you enjoyed this article, please keep in touch and sign-up to receive a free healthy living e-guide on josephinebila.com.

Quality of Care in Thalassaemia: Peer review

Dr Kate Ryan, Consultant Haematologist, Manchester Royal Infirmary UK

Dr Kate Ryan

As patients how can you be sure you are receiving the best up -to -date care?

A major challenge in England is that there is an uneven distribution of patients as this will depend on the parts of the country where people from groups at risk of thalassaemia settle. The prevalence of thalassaemia is highest in North and East London, West Midlands, North West England and Yorkshire. It is in these places that specialist teams and services for patients have developed.

However, patients who live outside these areas need to be able to rely on their local hospital as it is not possible to travel long distances to the specialist centre on a regular basis or if they acutely unwell. A survey 1999 showed that of just under half of 164 doctors caring for thalassaemia patients looked after a single patient only. In these circumstances patients may feel isolated and unsure of their service and it is difficult for the doctors and nurses to maintain expertise.

In recent years there have been a number of significant developments in the management of people affected by thalassaemia. These include MRI techniques to more accurately assess body iron loading which helps guide the best use of iron chelation drugs. These tests are expensive to set up and run and require expert interpretation if they are to be used in the best way. As a result they may not be available local to where patients live.

Following the introduction of universal newborn screening for sickle cell disease (SCD) in England (which also identifies most babies with Thalassaemia Major), it was recognized that the NHS needed to ensure the best follow on care for that baby was born and that this care needed to be lifelong.

There have been a number of approaches to improving the quality of care for people with haemoglobin disorders to ensure good care is available to everyone, regardless of where they live.

Standards of care

The UK Thalassaemia Society led the way by developing Standards of Care detailing not only the medical approach, but what patients could expect from services and the support they should receive (first in 2005, updated in 2008).

The National Haemoglobinopathy Registry (NHR)

This is a confidential database which hopes to collect information on the prevalence and treatments given for people with Thalassaemia and SCD in England. This valuable resource allows for planning of services and provides a way of seeing how many patients are getting the recommended tests, expert reviews and the types of treatment they are receiving.

National Specialist commissioning

NHS Commissioners in England are responsible for paying for the care that hospitals provide. They have a responsibility to make sure that each patient receives consistent care in line with best practice and that there are no major geographical variations depending on where you live. Thalassaemia and sickle have been recognized as needing a national approach due to the specialist nature of the condition and the variation of experience around the country. A model is proposed whereby a specialist haemoglobinopathy centre (SHC) who have the necessary expertise and resources link up with surrounding hospitals caring for all the patients who live in that geographical area. The SHC will have a lead responsibility for the care of all patients and will agree shared-care arrangements so that the patient can receive care locally, if appropriate. They will ensure that all patients have an expert annual review which, for thalassaemia patients on transfusion, will include MRI scans and other tests to monitor iron. They will also lead on training of staff dealing with haemoglobinopathy patients.

Peer review of Haemoglobinopathy services

Peer review is a process whereby teams review the care given by their colleagues in other hospitals using defined quality standards. The first round of peer review for Children and Adults with haemoglobinopathies has now been completed.

For haemoglobinopathies, the standards were based on the UKTS and sickle standards and covered information and support for patients as well as medical and nursing care and ways in which services monitored how well they were doing. Users of the service were involved in the setting of the standards and as reviewers on the visits. Review teams of doctors, nurses, users/carers and managers visited hospitals and gave feedback on how well they met the standards and things they might consider improving. A very important part of the process was speaking to patients at the time of visit to get their direct feedback on how they felt about their care.

The visits confirmed that services do vary considerably across England. Most

teams however provide a reasonable standard of care despite working in difficult circumstances with low levels of staff. Feedback from the users of services was generally positive for the specialist teams but they still had concerns about other aspects of care e.g. treatment in the A&E departments and lack of transfusions at convenient times. Notable areas for improvement included the lack of staff to deliver crucial aspects of care including community and psycho-social support. Information on patient outcomes was lacking and most teams knew of patients who were not attending for specialist review. There were very few networks of care but it is hoped these will strengthen over the next few years.

The next round of peer review is in the planning stage and it is proposed that the new standards will build on the findings of the last round and will aim to focus more on quality measures such as audit, patient satisfaction and the effectiveness of care.

The peer review steering group is always very keen to involve users in the peer review process as they are often the best ones to judge how the teams care for their patients. Anybody wishing to get involved should contact Sue McIldowie at sue.mcildowie@nhs.net 0121 507 2891 or 07854 387 186

Detail of the peer review process, the quality standards and reports of the findings can be accessed at www.wmqrs.nhs.uk.

Valuable research shows that it is possible to effectively screen for thalassaemia major in newborns

As many of our readers will be aware, all babies born in the UK have a sample of blood usually collected by a midwife at around 5 days after birth. The midwife collects the sample by pricking the baby's heel with a special device and collecting a few drops of blood on a special card. This is done to test the baby for a number of genetically transmitted conditions, for example sickle cell disease and cystic fibrosis. Although it is possible to detect significant thalassaemias from the bloodspot test, to date the UK National Screening Committee has not recommended screening for thalassaemia. However, it formally supports the current practice of reporting clinically significant thalassaemias found as a by-product of newborn screening for sickle cell disease - a position which UKTS finds difficult to understand. We know that there are between 20-30 babies born with significant thalassaemias every year; and there can be no doubt that early diagnosis is a benefit to these babies and their families - as the baby can be immediately referred to a haematologist for monitoring so that transfusions can be initiated at the appropriate time. This recently published paper shows that it is possible to effectively screen for thalassaemia major in newborns; and in practice laboratories have for some years report their findings if they indicate that the baby has a significant thalassaemia. This being the case, UKTS does not see any valid reason why thalassaemia major should not be officially included in the newborn screening programme. We will continue to lobby the National Screening Committee to officially include thalassaemia in newborn screening at every opportunity.

Newborn bloodspot results: predictive value of screen positive test for thalassaemia major

Allison Streetly, Radoslav Latinovic, Joan Henthorn, Yvonne Daniel, Elizabeth Dormandy, Phil Darbyshire, Debbie Mantio, Laura Fraser, Lisa Farrar, Andrew Will and Lesley Tetlow

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The online version of this article can be found at:

http://msc.sagepub.com/content/early/2013/11/25/0969141313514217

Abstract

Aim: There are limited published data on the performance of the percentage of haemoglobin A (Hb A) as a screening test for beta thalassaemia major in the newborn period. This paper aims to analyse data derived from a national newborn bloodspot screening programme for sickle cell disease on the performance of haemoglobin A (Hb A) as a screening test for beta thalassaemia major in the newborn period.

Methods:

Newborn bloodspot sickle cell screening data from 2,288,008 babies were analysed. Data reported to the NHS Sickle Cell and Thalassaemia Screening Programme in England for the period 2005 to 2012 were also reviewed to identify any missed cases (4,599,849 babies).

Results:

Within the cohort of 2,288,008 births, 170 babies were identified as screen positive for beta thalassaemia major using a cut-point of 1.5% HbA. There were 51 identified through look-back methods and 119 prospectively identified from 4 screening laboratories. Among 119 babies with prospective data, 7 were lost to follow up and 15 were false positive results. Using a cutoff value of 1.5% Hb A as a percentage of the total haemoglobin as a screening test for beta thalassaemia major in the newborn provides an estimated sensitivity of 99% (from the look back arm of the study) with a positive predictive value of 87% (from the prospective arm of the study). Excluding infants born before 32 weeks gestation, the positive predictive value rose to 95%.

Conclusion:

A haemoglobin A value of less than 1.5% is a reliable screening test for beta thalassaemia major in the newborn period.

From Trinidad to London – my journey so far

by Roanna Maharaj

Based on her achievements to date, Roanna was chosen to represent Trinidad and Tobago as a Student Ambassador. Here she is meeting the Queen at the Commonwealth Reception, Buckingham Palace, 28th October 2013.

Like many of the others, I was diagnosed with thalassaemia at six months of age. I was actually born in the sunny Caribbean island of Trinidad in 1989 where, back then, thalassaemia was virtually unheard off. Once diagnosed, my parents were advised by the doctors to abandon me in the hospital and have another child. The doctors thought they were doing the best for everyone, as to them thalassaemics did not live past the age of 16 anyway. What they did not know about my mother however, was that she did not recognise some words- "No, cannot, impossible....to list a few". She was determined to prove them wrong by firstly trying to ensure that I had the best possible treatment available and also striving for the "cure". She also tried to ensure that I benefitted from a "normal" upbringing.

Now despite having a positive upbringing, I never planned for a future as the constant grief I experienced each time one of my friends died really had me awaiting my turn…As I got older, I counted the years as deep down I really felt I was going to die like the others- so school was never a serious part of my life.

Life in Trinidad was a challenge as I remember many parents having to wonder every month where their child's next unit of blood would come from. You see in the Caribbean, the blood situation is very different to the United Kingdom as in order to receive blood, you have to donate- it really works like a cash machine- you have to deposit to in order to collect. I was quite fortunate as my parents always had donors for me but not all families shared my luck. For example, when I was growing up, there was this family with two thalassaemic children. On one occasion, I remember the parents were given one unit of blood and told to decide which child needed it most. I mean can you imagine being in that position? It was horrible.

In some hospitals, patients were also given whole blood (containing white cells and everything else). When questioned, the doctors treating those patients explained that they were trying their just to keep the patients happy as sooner or later they would die anyway and it was the faster and cheaper option for the hospital.

Well imagine, if getting something easy as blood was a problem, what about chelation? Desferal was the only chelator approved and not always available. This was also a major problem as even in 2004, patients were still having desferal through intramuscular injections instead of subcutaneous.

Now I bet after all that you wouldn't question why my mom and I left sunny Trinidad to relocate to rainy London right? Well, one of our main motivations for moving here, stemmed from attending our first international conference in 2002. It was the first time I had ever seen an old patient, well one that was a lot older than me (and he probably would be mad at me for saying this but it was George Constantinou.) I must say I was really inspired as George was there proudly showing off pictures of his beautiful daughter Iliya. I remember thinking to myself ....wait he's old and he has a family? So we can live past 16?

That conference also provided information on specialised testing and chelation therapies available in the UK which my mother immediately acted upon. Although we were advised against it and assured by my doctor in Trinidad that I was extremely healthy and my iron levels were perfect, my mother booked an appointment for me to have the T2* testing and a consultation with Dr Wonke at the Whittington Hospital. This was my first eye opener as not only did she speak to me directly, which was a first for me, Dr Wonke also told me that I was a "walking time bomb" since she discovered that both my liver and heart were severely iron overloaded. Based on her recommendations for immediate combination therapy, which was not available back home, our only option was to relocate to the United Kingdom.

Now despite my deepest worries about dying, on moving to London, I was presently surprised. It was an amazing experience, to walk into the thalassaemia unit and not be the oldest patient, but one of the youngest. I met older patients like Chris, Andy, Michael and the late Costas who continued to inspire me. I then started thinking about my future, you know about university and even about getting married someday.

My biggest challenge was choosing what clothes to wear for my transfusion which to me was more of a social event. I no longer had to worry where my next unit of blood came from, whether I would be over transfused or whether tap water would be used to flush my veins. Don't get me wrong though, I am not complaining, I do value the doctors enthusiasm for efficiency, as in one instance, two units of blood were simultaneously transfused, one unit in each hand- in order to shorten my stay at the hospital.

Despite being in the UK and having the best possible treatment, I started becoming one of those rebellious teenagers who didn't want to do my chelation. It wasn't because I was being stubborn (well maybe a little), it was because my ferritin didn't really affect me. You know, I didn't feel sick so I didn't see the point of taking anything. I started thinking that I could put it off for tomorrow, or the next day" and you know it just kept going on. I would forget to take my tablets; I would even break the needle off the thalaset, to make it look like I actually had the pump on when really I didn't just so that my mom wouldn't know. Yes parents, be aware of all the tricks of the trade!

However, my perception of chelation changed when during some time spent in Canada. In Canada, treatment was also not the best- they weren't as fortunate as the UK to have all three chelators available. During one of my visits to the clinic, I met a patient and I must say, she was the reason I turned my life around. This patient, who was finding it hard to take desferal, had been diagnosed with heart failure and diabetes and really wanted to be on the Ferriprox which was known to be the best chelator for the heart. After seeing her crying and knowing that I wasn't far away from her position, I then realised just how lucky I was.

In the UK, we are fortunate to have access to the proper care and treatment options, I was also able to prevent developing complications. I mean taking the pump and my ferriprox seemed a lot easier to me than injecting myself with insulin three times a day- or worrying whether I would ever be able to play sports again. To be honest, if my health deteriorated, it wasn't my parents fault or my doctors' fault for not taking care of me- it would actually be my fault. I am responsible for my own health. I must say though, after my little self realisation moment- and doing my treatment as prescribed (and that wasn't one today, one next month) my ferritin decreased drastically from 6000 to 471. In addition, there was no iron loading in my liver or my heart.

The truth is yes we have thalassaemia, but if we take care of ourselves and not make excuses, we are no different to anyone. We won't be classed as disabled but differently abled! We have the potential to do and be whatever we want- as now with the right treatment there is no stopping us.

Remember the little girl who once believed she was going to die at age16, well she is now 24 doing her PhD in psychology.

Meet Millie Hayes,
one of our young "thalassaemia family" members from North of the border. Millie and her classmates were celebrating St Andrew's day (30th November) by wearing tartan dresses to school on Friday 29th November. Millie lives in Glasgow and is under the care of Dr Elizabeth Chalmers at Yorkhill Hospital.

Imam Hussain Blood Donation Campaign

By Sarah Jawad, Marketing Co-Director 2013

NHSBT staff and campaign volunteers, East London, December 2012

As public health campaigns evolve and the focus shifts to education and awareness, so does the target audience. There is now an increased emphasis on campaigns which target the youth, with the belief that if an attitude is engendered from as early as possible, it will continue to evolve with a person. Recently, there has been a greater deal of attention given to the topics of blood and organ donation. The NHS has launched multiple advertising campaigns in an effort to engage with the youth and with parts of the UK population where blood donation is less common. The number of blood donors has been on the rise, as the number of donation centres rises and awareness is increased as to the conditions which need this life saving action.

There is, however, a worrying lack of both awareness and donations amongst the ever-growing ethnic minority population of Britain. We are a multicultural society, and with that comes a mixed gene pool, and an increasing prevalence of certain disorders. Amongst these are blood disorders more common amongst certain ethnic minorities, such as sickle cell anaemia amongst African-Americans, and thalassemia amongst Middle-Eastern and Mediterranean populations. Thankfully, our treatment approaches have developed, and modern medicine aims to control, if not treat, such conditions as much as possible. However, such blood disorders as thalassemia require multiple transfusions as part of the treatment plans and efforts to help normalise the lives of sufferers as much as possible. With an increased prevalence amongst ethnic minorities, it stands to reason that the blood most likely to be compatible with the majority of sufferers would, based on genetics, come from ethnic minority donors. A mismatch of blood types can lead to extremely serious medical complications, exacerbating what is already difficult time for patients. As such, blood banks need to be prepared with blood types compatible for those who need it most, such as thalassemia sufferers. Therein, however, lies the problem.

The NHS needs about 7,000 voluntary donations of blood per day. Currently. only 4% of the UK population are donors. This does amount to an impressive 2.5 million donors, but let's do the math. If every one of these enlisted donors donated at least once per year, that would amount to barely covering current NHS requirements for a single year. Add some reality into it - many donors do so on a one-off basis, and some blood types are more common than others, resulting in an unequal distribution of blood types amongst the donations. The result? A deficit in the amount of blood needed by the NHS. A deficit in the number of lives that can be saved.

In a population of 65 million, a fraction are donating blood. Of that fraction, less than 3% of them are from ethnic minorities. This exacerbates the already worrying deficit in available blood compatible for those from other ethnic backgrounds. The exact reason for this is unknown, though there are many possibilities - lack of awareness, poor education on the topic, poor communication and outreach to communities who traditionally do not have effective public health systems in place. What is known, however, is that something can be done about. Several proactive movements have risen up across the UK, sprung from the concerns within

NHSBT staff and campaign volunteers, East London, December 2012

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different ethnic communities as to the lack of education about blood donation and actual donors within their community. The Imam Hussain Blood Donation Campaign is one such example of this.

Established in 2006, the Imam Hussain Blood Donation Campaign seeks to increase the number of blood donors from Muslim communities, whilst at the same time raising awareness and providing education on the importance of blood donation and the conditions which require regular supplies of blood, such as thalassemia. Several members of the Muslim communities targeted by the campaign come from Middle-Eastern and Mediterranean countries with a high prevalence of thalassemia, and so it is a topic which is especially dear to the campaign's hearts. The campaign runs annually during the month of Muharram of the Islamic calendar, in conjunction with NHS Blood and Transplant, and now has established donor centres across the UK. Numerous cities, from Scotland to Surrey, work together to set up donor centres and encourage members of the community to donate blood, and the campaign has continued to grow with each year.

The name of the campaign was inspired by Hussain, the grandson of the Prophet Muhammad (peace be upon him), a figure celebrated in Islam for standing up for values which was universal in their significance, and making a stand against oppression and tyranny, and for justice and equality. The example he set of the ultimate sacrifice thus inspires Muslims to give back as much as they can to a community. However, the campaign may be thus named, but the aim is far more expansive. The Campaign aims to impress a crucial message to as wide an audience as possible, which is that there is a dire need for blood donors in this country, and even more so for blood donors from ethnic minorities. With every blood donation, 3 adult lives can be saved. There is so little to lose by taking part, and so much to gain, and it is both refreshing and inspiring to see the number of proactive movements encouraging donation, and societies such as the Thalassemia society, devoted to raising awareness of these conditions and aiding as much as possible. If anyone would like to know more about working in their community or how they can help, the Imam Hussain Blood Donation Campaign takes a universal approach, and can be contacted on giveblood@ius.org.uk. We look forward to such campaigns increasing in size and number with each coming year!

Archbishop of York retires from NHS Sickle Cell & Thalassaemia Screening Programme

It is with regret that we report that The Most Reverend and Right Honourable the Lord Archbishop of York Dr John Sentamu has announced his retirement from his position as Lay Chair of the Steering Group of the NHS Sickle cell and Thalassaemia Screening Programme. We are grateful to the Archbishop for the years he has chaired the Steering Group; and we know that he will continue to act as a champion for the cause of thalassaemia and sickle cell disease in the House Of Lords. The Archbishop is seen here at his final meeting on 7th November 2013 (Hughes Parry Hall, Central London) with Cathy Coppinger, the Programme Manager.

Recent Events & Meetings

Those who attended meeting on behalf of the UK Thalassaemia Society are: Gabriel Theophanous President, Mike Michael Vice-President, Romaine Maharaj Treasurer, George Constantinou Secretary, Andy Charalambous Asst. Secretary, Elaine Miller National Coordinator, Katerina Loizi-Read Office Administrator, Roanna Maharaj patient member

Acronyms

APPG	– All Party Parliamentary Group for Sickle Cell & Thalassaemia
NHSBT	– NHS Blood & Transplant
TIF	– Thalassaemia International Federation
UKFHD	– UK Forum on Haemoglobin Disorders

- APPG – All Party Parliamentary Group for Sickle Cell & Thalassaemia
- NHSBT – NHS Blood & Transplant
- SCTSP – NHS Sickle Cell & Thalassaemia Screening Programme
- TIF – Thalassaemia International Federation
- UKFHD – UK Forum on Haemoglobin Disorders
- 7 September 2013 – awareness presentation, Islamic Unity Society conference, Hammersmith, London W6 *Elaine Miller*
- 13 September 2013 – awareness presentation, Central London Sickle Cell & Thalassaemia Services conference *Elaine Miller*
- 4 October 2013 – steering group meeting, research project on the social implications of being a carrier of unusual haemoglobins, University of York *Elaine Miller*
- 12 October 2013 – awareness presentation, Newham Sickle Cell & Thalassaemia Services conference, West Ham FC *Elaine Miller*
- 15 October 2013 – awareness presentation, Sheffield Chinese Community Centre *Elaine Miller*
- 20-14 October 2013 – TIF International Conference, Abu Dhabi – Gabriel Theophanous, Mike Michael, George Constantinou, Andy Charalambous, *Romaine Maharaj*
- 22 October 2013 – Sheffield Sickle Cell & Thalassaemia Foundation conference, Sheffield *Elaine Miller*
- 27 October 2013 – awareness presentation, Manchester Sickle Cell & Thalassaemia Service 30th anniversary conference, Marriott Hotel Manchester *Elaine Miller*
- 31 October 2013 – meeting with NHSBT Yorkshire & Humber, Northern General Hospital *Elaine Miller*
- 6 November 2013 – awareness presentation, GP surgery North London Katerina *Loizi-Read*
- 7 November 2013 – Steering group meeting, NHS Sickle Cell & Thalassaemia Screening Programme, Hughes Parry Hall, Central London George Constantinou, *Elaine Miller*
- 7 November 2013 – awareness presentation, GP surgery North London Katerina *Loizi-Read*
- 21 November 2013 – UK Forum on Haemoglobin Disorders academic meeting, Nottingham *Elaine Miller*
- 25 November 2013 – All Party Parliamentary Group for Sickle Cell & Thalassaemia meeting, Portcullis House, Westminster *George Constantinou, Roanna Maharaj*
- 2 December 2013 – Hepatitis C Expert Policy Group, King's Cross, London *George Constantinou*
- 3 December 2013 – awareness presentation, University of Northampton *Elaine Miller*
- 5 December 2013 – awareness presentation, Sheffield Hallam University *Elaine Miller*
- 6 December 2013 – health awareness day, Central Hall, Keighley, West Yorkshire *Elaine Miller*

Help us to help you by supporting YOUR Society – every £1 is precious!

Please Support The UK Thalassaemia Society by Making a Monthly Donation

STANDING ORDER FORM

To the Manager *[Name of Your Bank]*
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Please pay: *Bank of Cyprus UK, PO Box 17484, 87 Chase Side, London N14 5WH*

For the credit of: *UK Thalassaemia Society, Registered Charity No: 275107*
Sort Code 30-00-42 Account Number 00593812

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

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

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UK Thalassaemia Society, 19 The Broadway, Southgate Circus, London N14 6PH.
We will then send this form on to your bank.

Thank you for your valued support.

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UK Thalassaemia Society, 19 The Broadway, London N14 6PH
Charity Reg No. 275107

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

Ethnic Origin: *(Optional)*

Contact Details

Telephone: *Home:*

Work:

Mobile:

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Are you a:

☐ Patient ☐ Parent/Relative
☐ Healthcare Professional ☐ Association
☐ Other (Please state)

Membership Required *(please tick)*

☐ ANNUAL **(£10.00)** ☐ LIFE **(£100.00)** *(Please make your cheque payable to **U.K.T. Society**)*

If you are a patient or parent of a patient please complete the section below

Patient's Name(s):

Date of Birth:

Sex: ☐ Male ☐ Female

Type of thalassaemia: *(e.g. Major, Intermedia, Haemoglobin H etc)*

Hospital where-treated:

Address:

Consultant's Name:

Consultant's Telephone:

GP's Name:

Address:

Telephone:

Blood Transfused *(please tick)*

☐ Whole ☐ Washed ☐ Frozen ☐ Filtered

Chelation *(please tick)*

☐ Desferal ☐ Deferiprone ☐ Desferal & Deferiprone

Transfusion Frequency:

Units received at each transfusion

Blood Type

OFFICE USE: Date Paid _____ Receipt No._____ Approval Date _____

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