HCC
South East London and South East

South Thames Sickle Cell & Thalassaemia Network

Issue 18

RedCell Newsletter

Welcome to the latest edition of Red Cell News Issue 18.

Despite the challenges of 2021/22, there have been many positive developments across the South East London and South East region. There have been various educational events to provide staff from a wide range of clinical backgrounds and at different hospitals with the opportunity to gain further insight into sickle cell, thalassaemia and rare anaemias.

We look forward to our annual Sickle Cell Awareness educational patient day which is due to be held on the 13th July virtually via Zoom.

Our local support groups have continued to meet virtually on a monthly basis to keep patients informed about the latest topics of interest and provide an informal opportunity to meet with other patients and staff.

There have been a number of new developments including the introduction of new treatments such as Crizanlizumab and Voxelotor; stem cell transplants at King's for adults with sickle cell disease from across the region; and changes proposed in response to the 'No One's Listening' All-Party Parliamentary Group report into Sickle Cell and Thalassaemia management following inquiries into avoidable deaths and failures of care for sickle cell patients in the NHS.

We have new starters who we welcome to our network, as well as some members of our team who we wish all the best as they leave to pursue other challenges. Professor Jo Howard leaves our team having made enormous contributions to sickle cell disease care both locally and nationally, including as the lead of our South East London and South East Haemoglobinopathy Coordinating Centre. She leaves us to become part of a group aiming to achieve successful gene therapy for sickle cell disease and thalassaemia. We are all immensely grateful for her efforts over the years and wish her great success in her next venture.

We continue to update our website to include information for patients and clinicians, please take a look at www.ststn.co.uk

Mission Statement: "to improve patient experience by offering better treatment outcomes for people with sickle cell and thalassaemia"

Visit **patient zone** on our website: **www.ststn.co.uk** to find patient information, clinic times, support group information and previous editions of the red cell newsletters.

KING'S HEALTH PARTNERS

Pioneering better health for all

Welcome Our New Welfare Advsior

My name is Richard Conway, and I am the new Welfare Support Advisor under Guy's and St Thomas' Sickle Cell and Thalassaemia Community Services.

I can provide general welfare advice for the network, and additional welfare case work support for residents of Lambeth, Lewisham and Southwark.

I have previously worked for:

* Support Through Court (RCJ) family courts in Holborn
* Kindship (Formally known as Grandparents Plus)
* MS Society
* Disability Law Services (Managed welfare helpline - training

WHAT SERVICES CAN I PROVIDE?

Welfare Benefits:

* Legacy - Employment & Support Allowance/ Housing Benefit/Income Support/Tax Credits System
* Universal Credit
* Personal Independence Payment/Disability Living Allowance/Attendance Allowance/Carers Allowance
* Council Tax

All other public funding UK welfare benefits. This also includes appeals and tribunal services

Employment:

* CV's
* Access to work
* Reasonable adjustment in the workplace

Including appeals and tribunal representation or referrals for legal aid, or other professional legal representation

Education:

* Student Finance
* Disability Student Allowance (DSA)
* Educational Healthcare Plans (ECHP)

Including appeals and tribunal representation or referrals for legal aid, IPSEA, SENDIAS, or other professional legal representation

Housing:

* Homelessness
* Disrepair
* Adaptations
* Rent Arrears

Including appeals and tribunal representation or referrals for legal aid, or other professional legal representation

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Richard Conway

Lewisham & Greenwich Psychology Service

Lewisham and Greenwich NHS Trust have recently established a Psychology Service for Children and Young People (CYP) and Adults with Sickle Cell & Thalassaemia. The service covers both University Hospital Lewisham (UHL) and Queen Elizabeth Hospital (QE) and was set up in June 2021 by the service lead, Gary Bridges, Principal Counselling Psychologist.

Prior to taking up this post, Gary worked in similar roles with adults with sickle cell disease and thalassaemia at King's College Hospital from 2017 to 2021, and at Homerton University Hospital in Hackney from 2015 to 2017. Gary is accredited as a Cognitive Behavioural Therapist and Interpersonal Psychotherapist, and has extensive experience of working with people with a diverse range of psychological and physical health problems.

Dr Joanna Adebayo is the Paediatric Clinical Psychologist for the CYP service, taking up her post in November 2021. Joanna recently obtained her Doctorate in Clinical Psychology at the Salomons Institute for Applied Psychology in 2021. Joanna has had prior experience working with children and young people in both mental health and paediatric services, and feels privileged to have the opportunity to contribute to influencing psychological care for children and young people with sickle cell disease and thalassaemia, and their families.

Dr Hannah Grocott is the Clinical Psychologist for the Adult service, taking up post in November 2021. Hannah completed her Doctorate in Clinical Psychology with the University of East Anglia in 2019. Prior to joining Lewisham and Greenwich Trust she worked as a Clinical Psychologist in an Early Intervention in Psychosis service. Hannah first became interested in sickle cell disease and thalassaemia in 2010 when she worked as an Assistant Psychologist at the Brent Sickle Cell and Thalassaemia Centre, Central Middlesex Hospital. Finding this an incredibly meaningful and often under-served area of physical health, Hannah was grateful for the opportunity to return to haemoglobinopathies and contribute towards the development of a psychology service to support the patients of Lewisham and Greenwich NHS Trust.

Welcome Our New Psychologist At KCH

My name is Giuliano and I am a HCPC registered Psychologist, a BABCP accredited Cognitive Behavioural Therapist (CBT) and an IPTUK accredited Interpersonal Psychotherapist (IPT). I have been training and working in the fields of psychology and psychotherapy for more than fifteen years, and I have professional experience in a variety of clinical settings and in different countries.

Since 2017 I have been working as a psychotherapist, with a particular focus on the treatment of depression, anxiety, OCD, trauma, low self-esteem and interpersonal difficulties. From 2018 I have been working - as a psychotherapist and as a clinical psychologist - in the NHS, in three different Trusts.

I strongly believe in change and in the fact that people can make positive steps towards their personal and relational growth and their emotional wellbeing. As a therapist, I feel honoured to have the chance to support in this delicate process. My diverse professional roles have allowed me to appreciate the difficulties people may face in sharing negative emotions and difficult aspects of their lives. This valuable experience has given me the skills to work with evidence -based therapeutic techniques whilst building a therapeutic alliance based on trust, collaboration and human values.

My main goal as a therapist is to support my clients to build on their strengths to overcome emotional difficulties and life challenges, and to help them become their own therapist by learning skills and techniques that they can continue to practise after treatment.

Dr Giuliano Sorrentino

How Does Sickle Cell Impact Daily Life?

THE ASCQME QUESTIONNAIRES!

In SCD services patients are offered annual review appointments with the medical team to review their physical health and management of their SCD and any other health concerns they are experiencing. During these reviews patients are given the opportunity to have a review of the impact of SCD on their quality of life with a psychologist. SCD can impact a number of different areas of an individual's life including their emotional wellbeing, their relationships and their education/ occupation. If SCD is significantly impacting an individual's quality of life in one or more areas, a psychologist may be able to help them to manage these difficulties and improve their wellbeing.

Psychologists often use questionnaires or measures to identify which area of people's lives are difficult and to what degree this causes them concern or distress. The psychologist will then ask the individual about their answers to understand their experiences and perspective so that they can think together about what kind of support would be most helpful. Most questionnaires that measure quality of life for individuals with a long-term health condition have been created using the experiences and feedback of individuals with a variety of health concerns. However, this has typically not included individuals with SCD. This can mean they do not address the very specific concerns and experiences someone with SCD may have.

The ASCQME questionnaires were created by a research team in America in 2019 using the experiences and feedback of a large cohort of people with SCD to ensure that the questions focus on the most relevant concerns that SCD patients have. These are the first and only questionnaires designed to measure the impact of SCD on peoples' daily lives. The ASCQME questionnaires include 7 different sets of questionnaires covering a range of issues including the impact of pain, emotional wellbeing, social functioning and sleep. The creators of the measures also found that some existing questionnaires addressing fatigue and thinking and memory abilities were also relevant for people with SCD.

Since November 2021 the GSTT Psychology team have trialled using these measures in face-to-face annual review clinic consultations to better understand the needs of our patients and help us to offer the most useful support to each individual seen in clinic. We are the first service in the UK to use these questionnaires and are excited to see that staff and patients are finding them helpful. ASCME is also being used in a large scale cohort study of individuals with SCD 'Natural History and Clinical Outcomes of Patients with Sickle Cell Disease' currently recruiting at GSTT and KCH. This will allow professionals to better understand the concerns and needs of people with SCD in various services.

If you would like to know more about these questionnaires?

please visit the ASCQME website http://www.ascq-me.org/ or contact the GSTT Psychology team at hps@gstt.nhs.uk

Have you already had experience of completing these questionnaires at GSTT?

GSTT Psychology team would love to hear your thoughts on these questionnaires – please contact us at hps@gstt.nhs.uk

We would also be happy to offer advice to any SCD services who would like to use these measures in their clinics

Please contact us at hps@gstt.nhs.uk if you would like to discuss this further.

Dr Abbie Wickham

Clinical Psychologist

Haematology Health Psychology Service, GSTT

Novel Therapies For Sickle Cell

Introduction:

After many years when hydroxycarbamide or blood transfusion were virtually the only options for treatment of sickle cell disorder (SCD), we are now at the dawn of a new era of disease-modifying treatment.

Hundreds of people with SCD worldwide have volunteered to pioneer over the years by enrolling in clinical trials, helping clinicians understand how these new drugs work, and how they can best be used in SCD. One consistent finding is that there is no 'one size fits all' approach for SCD. The condition can present with very different symptoms and problems from person to person.

A little bit of background biology:

The two faces of sickle cell disorder

The two main aspects of sickle cell disorder are vaso-occlusion and anaemia. *Vaso-occlusion* refers to the actual 'sickling' process, where the red blood cells suddenly change into a rigid sickle shape and block the bloodstream. *Anaemia* relates to the lack of red blood cells in comparison with a non-affected person. Red blood cells are responsible for oxygen transport, and a reduced number of red blood cells makes it harder for the body to deliver oxygen to the tissues.

Vaso-occlusion causes tissue inflammation and infarction, leading to pain and ultimately to damage of the affected area (for example, avascular necrosis, AVN). Anaemia can lead to fatigue and lack of stamina. Also, anaemia increases the rate of blood flow through the body. In order to try to compensate for the lack of oxygen delivery due to a lower number of red cells, the heart can increase the volume that it pumps around per minute. However, this increased flow puts a huge demand on the heart and can cause sheer stress in the blood vessels of delicate organs like the kidneys and brain.

Traditionally, the approach for sickle cell disorder has always been fluids and pain relief. Pain relief will help pain symptoms of SCD, but does not target any of the mechanisms above. Disease-modifying treatment aims to prevent the sickling-related damage rather than just managing the resulting pain.

Why do red blood cells sickle?

Through a genetic variation, the adult bone marrow stem cells produce red blood cells with an unstable liquid inside (haemoglobin S). Under usual circumstances, the cells maintain a normal shape and transport oxygen in the same manner as healthy red cells. After certain triggers, the liquid suddenly becomes 'solid and spikey' and turns the cell rapidly into a sickle cell shape. The red cell instantly loses its ability to carry oxygen and becomes very vulnerable to destruction, unless it is rapidly re-exposed to high oxygen levels. Before birth, the stem cells do not yet produce haemoglobin S but a stable variant (haemoglobin F or foetal haemoglobin) instead. After birth, most production of haemoglobin F is switched off in favour of haemoglobin S.

What is the cause of the anaemia?

In people *without* SCD a common cause for anaemia is iron deficiency. The cause for anaemia in sickle cell patients is not related to iron, and taking iron does not help – taking iron can sometimes even be harmful.

Because of the unstable haemoglobin S, the lifespan of a red cell in someone with sickle cell disorder is much shorter than average. The bone marrow is generally working overtime to compensate for the increased loss of red cells, but cannot keep up with the demand. This process of early loss or destruction of red cells is called haemolysis.

The waste products of haemolysis are also a hallmark feature of sickle cell disorder and are responsible for jaundice. Also, the waste products are thought to be partially responsible for sickle cell-specific problems like leg ulcers and priapism

Treatment options:

One can identify a number of possible targets that can prevent sickle cell-related damage or events:

* Change the bone marrow stem cells so they produce less haemoglobin S – or none at all
* Replace the red blood cells that contain haemoglobin S with healthy red blood cells containing normal haemoglobin
* Stop sickled red blood cells from getting stuck in the blood vessels
* Stabilise the haemoglobin S in red cells so it can no longer sickle
* Prolong the lifespan of red cells containing haemoglobin S

A Bone Marrow or Stem Cell Transplantation is currently the only available treatment in the UK that provides a cure for SCD, by replacing the haemoglobin S-producing stem cells with stem cells from a donor that produce a stable/normal haemoglobin. Patients undergoing this treatment receive radiotherapy and immune suppression to allow the donor cells to settle in the bone marrow without their immune system rejecting the donor cells. There are a number of limitations to this treatment and at present, only people who have a so-called matched sibling donor are eligible. There are potential complications and the procedure may not be successful, so a stem cell transplant is generally only offered to more severely affected patients.

Gene therapy is a very interesting and promising future development. Rather than using someone else's stem cells, in gene therapy the stem cells of the patient are 'edited' in the laboratory so they no longer produce haemoglobin S -containing red cells. Stem cells are collected from the patient's bone marrow, and after the editing of the DNA/genes, the patient will receive chemotherapy to eradicate the remaining Sickle Cell Stem cells in the bone marrow before the new stem cells are returned. This treatment is still in the early stages of development. We expect that clinical trials will come to the UK in due course.

An (automated) exchange blood transfusion replaces approximately 70-80% of the red cells containing haemoglobin S in one 2-3 hour session. This treatment is often used in an emergency and can be life-saving in the event of a severe cascade of sickling in several organs including the lungs.

It can also be offered as a routine treatment to prevent sickle cell-related problems. However, the lifespan of healthy red blood cells is limited to a maximum of 8-12 weeks and for this reason people who have blood transfusion for prevention will need to have this treatment once every 4-8 weeks, otherwise the haemoglobin S-containing red cell will become the majority and the patient will start experiencing symptoms again. Blood transfusion, and in particular a top-up blood transfusion can also be used to ameliorate the symptoms of anaemia.

Hydroxycarbamide can also change the stem cells in the bone marrow. Through an unknown mechanism, haemoglobin S-producing stem cells partly switch back to producing haemoglobin F after 3-6 months of exposure to the drug. The more haemoglobin F, the less likely the cells are to become unstable and turn into sickle cells. As a result, there is less vaso-occlusion and destruction of red cells. This in turn means less pain and damage, and an improvement of anaemia. Hydroxycarbamide is available as capsules, tablets or as a liquid. The dose varies from person to person.

The new drug crizanlizumab leaves the production of haemoglobin S-containing red cells untouched, but targets the actual vaso-occlusion by reducing the stickiness of blood cells. Rather than getting stuck and causing inflammation and infarction, the sickled red blood cells continue to flow through the blood vessel and will mostly not haemolyse (fall apart). The benefits of crizanlizumab include fewer painful vaso-occlusive crises, and possibly a reduced risk of developing AVN. However, there is no effect on anaemia or haemolysis. Crizanlizumab is given via a 4-weekly intravenous drip and people will have to attend the haematology day unit for the infusion.

The new drug voxelotor is a haemoglobin S-stabiliser. Voxelotor also leaves the production of haemoglobin S-containing red cells untouched. After taking voxelotor, the drug enters the red cell and keeps the oxygen levels within the red cell higher. By doing so, the threshold for the red blood cell to turn into the sickle shape becomes higher. The main effect is a reduction of cells falling apart (haemolysis), resulting in an improvement in anaemia and a reduced production of waste products (jaundice). The effect on vaso-occlusion and sickle cell pain is limited. Voxelotor is a daily tablet and the average dose is 3 tablets once a day. Voxelotor is not yet licensed in the UK but approval is expected later this year. It has been used in the USA for a while already and some people in the UK have been able to join an 'early managed access programme'

Etavopivat is an experimental drug not yet licensed for use anywhere in the world. King's College Hospital has access to this drug via a clinical trial for sickle cell patients as well as thalassaemia patients. Etavopivat has two mechanisms of action: it stabilises haemoglobin S in a similar way to voxelotor by increasing the amount of oxygen kept in the red cell. Via a different mechanism, it also replenishes the energy batteries (ATP) in the red cells which gives them a prolonged life expectancy. The expected effects are both improvement of anaemia and a reduction of vaso-occlusive painful crises.

What Can I do?

Many people with SCD and relatives of people with SCD still believe that disease-modifying treatment for sickle cell disorder is harmful. In fact, the opposite is true. Many people suffering from long-term complications and severe pain have previously never been offered therapy, or declined therapy. Choices are often based on the wrong assumptions or on strongly held views by leading people in the community or on social media.

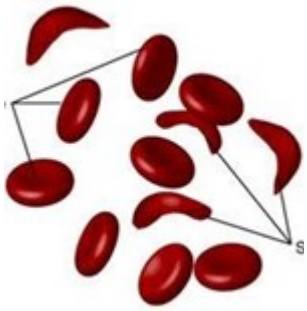
In this era, being well-informed and making sure that your health care providers listen to your needs is even more important than before. You can help by sharing your experience with others, and help others better understand their disorder and the available options.

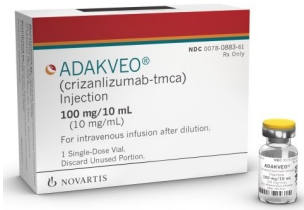
You can also help by contributing to clinical research. None of the treatments mentioned above would have become available without pioneers, people who volunteered to try a new drug. Either in the hope to benefit themselves, or because they want to contribute to the development of better healthcare for people with SCD.

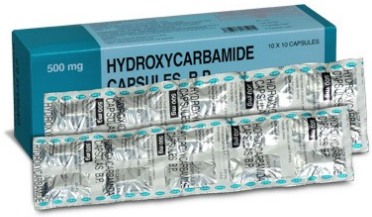
Clinical research does not necessarily involve trying a new drug like etavopivat; clinical research also involves sharing your health care record (anonymously) with researchers to help them better understand the natural course of sickle cell disorder ('Natural History and Clinical Outcomes of Patients with Sickle Cell Disease' is currently recruiting at GSTT and KCH).

Alternatively, you could for example participate in a project where extra scans are performed to help understand sickle cell-specific complications.

Your team is always willing to discuss treatment – old or new – and clinical trials with you.







Dr Arne de Kreuk
Consultant Haematologist
King's College Hospital

SPOTLIGHT ON THE NHP

National Haemoglobinopathy Panel

The NHP (UK National Haemoglobinopathy Panel) is a body that coordinates and oversees Haemoglobinopathy services in the UK, with an aim to improve quality across the nation. Though it is generally not known or seen by patients, most of its members are directly patient-facing and passionate about addressing the challenges that have been entrenched in the Haemoglobinopathy and Inherited Rare Anaemia communities, such challenges as geographically inequitable services, stagnant research and development, negative hospital cultures and their impact on patients with haemoglobin disorders, and general funding and manpower issues.

The NHP is the operational arm of the Clinical Reference Group (CRG) for Haemoglobinopathy Services, commissioned by NHS England and NHS Improvement (NHSEI) in 2019 along with the Haemoglobinopathy Coordinating Centres (HCC), which connect together and oversee Haemoglobinopathy services according to geographic regions.

The NHP, chaired by Professor Baba Inusa, with Professor John Porter as the Deputy Chair, is made up of experts from all the HCCs. The NHP works closely with King's Health Partners, South East London and South East HCC, CRG, Sickle Cell Society (SCS), UKTS (UK Thalassaemia Society), UK Forum for Haemoglobin Disorders (UKFHD) and others, to form the NHP network.

Access to the vast array of expertise of this network are the special element that fortifies the monthly MDTs (clinical case reviews), policy action, practice guidelines, research, and many other joint efforts.

Since its inception on 1st January 2020, the NHP's output has directly improved patient outcomes, as it liaises between its own network and NHSEI. Improvements include increased training for Haemoglobinopathy and non-specialist practitioners and patients, facilitating outreach clinics to support Haemoglobinopathy teams in need, creating and easing patient access to novel therapies and treatments (e.g. Crizanlizumab, Voxelotor and Stem Cell Transplantation for adult sickle cell disease), developing the National Haemoglobinopathy Registry (NHR), publishing treatment/policy guidelines, and amplifying the Haemoglobinopathy patients' plight and voice in parliamentary discussions, one of which led to an enquiry and, subsequently, the *No One's Listening* report. The NHP has coordinated the development of SOPs for Crizanlizumab and Voxelotor and consensus statements on paediatric stem cell transplant, PIMS-TS and strategic response to the COVID-19 pandemic.

The NHP network, despite many challenges past and ongoing, continues to champion the cause of Haemoglobinopathy and Rare Anaemia patients.

https://www.nationalhaempanel-nhs.net
twitter: @NatHaemPanel
Instagram: @nhp.uk
LinkedIn: www.linkedin.com/in/nhp-uk-nhs

World Sickle Cell Day 19th June 2022

UKTS Fun Run

United Kingdom Thalassaemia Society (UKTS) hosted its first Fun Run on Sunday May 8th 2022 at Grovelands Park. This event was initially planned as a local celebration of International Thalassaemia Day but was opened up to the wider community following requests from friends and families wanting to support from further afield.

The day was enjoyed by over 300 supporters in the park, with just over 100 signed up to run or walk. Globally, it was supported by over 30 countries with participants opting to wear red, complete the distance and provide pictures of themselves and their groups which was shared on social media platform, painting the various platforms red.

The event was supported by Peter Polycarpou and Kypros Kyprianou, patrons of the society, together with Bambos Charalambous, Member of Parliament for Enfield Southgate and Chair of the All Party Parliamentary Group for thalassaemia. They were all joined at the starting line by Michael Yiakoumi (Parikiaki) and Katerina Neocleous (LGR radio) together with the chair of the society Gabriel Theophanous and members of the society.

The commitment shown for the society was fantastic and the charity raised just over £12000.00 in donations, sponsorship and funds raised by our patrons and supporters.

The feedback received has also been very positive with many requests for similar events.

Awareness Corner

Manchester took the spotlight on Saturday May 21st 2022. Dr Sabiha Kausar and TeamUKTS provided information and literature to over 500 people at public event held in Altrincham.

There were lovely displays and the event was attended by a wide cross section of our community.

Special thanks to Dr Sabiha Kausar for organizing this event.

Another successful awareness event in Coventry. Nabeel Javed chose to fulfill his dream by entering the boxing ring to prove that you can accomplish anything despite having thalassaemia major. His message was received by over 600 attendees. Well done Nabeel!

Patient Event

Saying goodbye to Professor Jo Howard

Professor Jo Howard was a consultant haematologist and sickle cell service lead at Guy's and St Thomas' for 15 years.

She left her post in June 2022 to take up a new role with a large pharmaceutical company who are looking to develop curative treatments for sickle cell.

Prof Howard was very dear to her patients and the patient support group reflected this by organising an excellent farewell for her.

A number of her patients sent in written and recorded video messages of appreciation which were played on the day. Some patients gave speeches in person.

The group also organised a hamper as a token of thanks which was presented to Prof Howard at the event on 21st April 2022.

There was excellent attendance from her patients and colleagues, and it was an afternoon of appreciation filled with memories, laughter and tears.

Patient Zone

PAIN MANAGEMENT TIPS!

Try a massage, acupuncture or relaxation techniques!

Use a heat pad or take a warm bath!

Drink water or other fluids when your symptoms start. Staying hydrated can help you head off the worst of an attack!

Do you have a story to tell?

Email info@ststn.co.uk If you'd like to share your experiences or would like to contribute to red cell news in any way.

The network is proud to announce that we now have a YouTube channel (STSTN) which has plenty of content including our network educational events.

Please don't forget to:

LIKE, COMMENTS, SHARE & SUBSCRIBE.