KING'S HEALTH PARTNERS
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South Thames Sickle Cell & Thalassaemia Network

red cell news

A tri-annual newsletter for patients with sickle cell disease and thalassaemia

Issue 2 Winter 2012

ABOUT STSTN

The South Thames Sickle Cell and Thalassaemia Network (STSTN) is a new haemoglobinopathy collaboration led by Consultant Haematologists and Paediatricians at King's College Hospital, Evelina Children's Hospital and Guy's and St. Thomas' Hospital NHS Foundation Trusts.

STSTN aims to:

"improve the patient experience by offering better treatment outcomes for people with sickle cell disease and thalassaemia"

Find out more at: www.ststn.co.uk

IN THIS ISSUE

WELCOME TO THE 2ND EDITION OF RED CELL NEWS

NETWORK NEWS

PATIENT FORUM DAYS

Patient Forum Days were held around the Network in July at Guy's & St. Thomas' and King's College Hospitals. 73 patients and families joined staff at Guy's and St. Thomas' Hospital on 12 July for their annual Sickle Cell Awareness Day.

These events give patients and their families an opportunity to learn more about the services on offer at each hospital, to meet staff and to provide information on how best to manage their condition.

Dr Nicky Thomas, Consultant Health Psychologist at Guy's and St. Thomas', said: "The event provided a great opportunity to communicate information about service developments, and research as well as providing a forum for patients to give their views about their experience of the service."

More on this story can be found on page 3

CHILDREN'S CHRISTMAS PARTY AT THE EVELINA CHILDREN'S HOSPITAL

Evelina Children's Hospital will be holding a Christmas Party on the afternoon of Tuesday 18th December and all of our sickle cell children are invited! Please contact Florence on florence.bristow@gstt.nhs.uk or 0207 188 7774.

STSTN WISHES YOU A HAPPY CHRISTMAS AND NEW YEAR.

We hope you have a Happy Christmas and wish you all the best for the New Year.

Issue 3 will be out in Spring 2013. If you have a story to share, any comments or suggestions for the newsletter, events to publicise or you would like to offer your illustrating or photographic skills to red cell *news* please get in touch. We would love to hear from you.

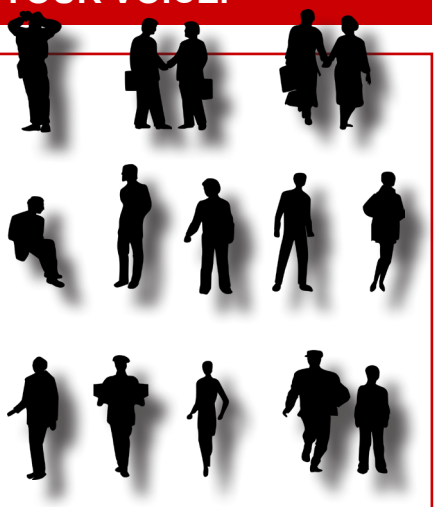
Until the Spring!

Annabelle
STSTN Support Manager
info@ststn.co.uk
020 7848 5441

PATIENT PLATFORM

YOUR STORIES. YOUR NEWS. YOUR VOICE.

GET IN TOUCH



Do you have a personal story about your experience with sickle cell disease or thalassaemia that you would like to share?

Is there an issue about sickle cell disease and thalassaemia that you would like to discuss?

Do you have an event you would like to publicise or would you like to connect with other patients and create a support group?

Or would you simply like to tell us what you think of red cell *news* or suggestions of what you would like to see included.

Then we would really like to hear from you.

Get in touch with Annabelle by email or phone:

info@ststn.co.uk or 020 7848 5441

NETWORK NEWS

6TH ANNUAL SICKLE CELL AND THALASSAEMIA ADVANCED CONFERENCE, 24-27 SEPTEMBER 2012

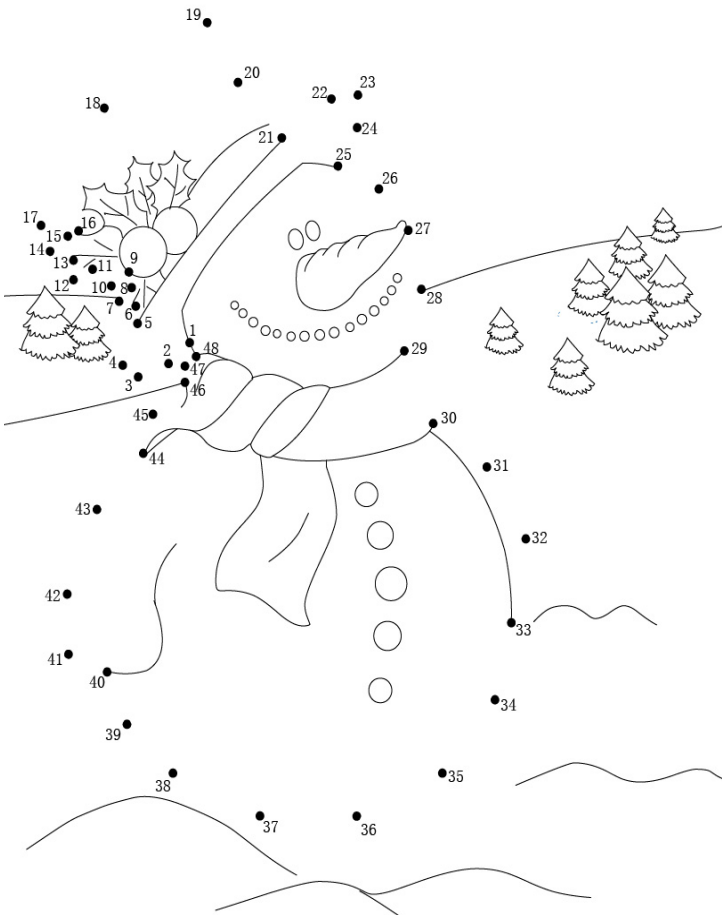
Guy's & St. Thomas' Hospital hosted the 6th Annual Sickle Cell and Thalassaemia Advanced Conference in September, with cutting edge presentations by a number of the world's leading experts in sickle cell and thalassaemia.

Chairman of Guy's & St. Thomas', Sir Hugh Taylor, opened the conference, which highlighted opportunities for developed countries, including the UK, to influence global research.

There was also an emphasis on the benefits of universal screening for Sickle Cell and Thalassaemia and the need for comprehensive follow up programmes. The Sickle Cell Research (SCORE) team presented a video of their work in Nigeria.

continued on page 3

KID'S CORNER - JOIN THE DOTS AND COLOUR IN...



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The conference was attended by 100 professionals from across the globe. We would like to thank Novartis for supporting the conference with an unconditional educational grant, Lois Whittaker and Ian Mackie and their team at GSTT for organising the event, and our esteemed colleagues who chaired sessions throughout the conference. The next course will take place on 3 - 5 October 2013.

Delegates at the 6th Annual Sickle Cell & Thalassaemia Conference

SICKLE CELL IN FOCUS, 24 - 25 MAY 2012

King's College Hospital and King's College London hosted another hugely successful Sickle Cell in Focus conference in May. Now in it's 6th year, this two-day intensive, educational conference had delegates and speakers from the UK, Europe, America and the Middle East. Topics included updates on reasearch, and management for complications in sickle cell diseaes and debates on controversial issues.

With over 100 guests, the conference continues to be a great success and we look forward to Sickle Cell in Focus 2013, taking place on 6 - 8 June.

Conferences like these help us to better understand and treat sickle cell disease, with the primary aim of making our patient's lives better.

PATIENT FORUM DAYS: SPREADING THE WORD ABOUT SICKLE CELL

Dr Nicky Thomas Consultant Health Psychologist, Guy's and St. Thomas' Hospital

Did you know that there are over 9,000 people living with sickle cell disease in London? Sickle cell disease is a serious disorder in which the body makes sickle (or crescent) shaped red blood cells. These cells tend to block blood flow in the blood vessels of the limbs and organs. Blocked blood flow can cause pain, serious infections and organ damage.

Neil Westerdale
Advanced Nurse Practioner

Sickle cell disease is the most prevalent genetic disorder worldwide and it primarily affects people of African and Caribbean origin as well as small numbers of people from the Mediterranean, Middle East and India.

July is sickle cell awareness month which aims to raise public understanding of the condition. There are around 700 adult and 450 paediatric patients with sickle cell disease at Guy's and St Thomas', with similar numbers at King's College Hospital and these numbers are predicted to rise. The adult teams at both hospitals held a Patient Awareness Day for patients, families and staff to educate and inform them further about managing their condition.

Guy's and St. Thomas' and King's College Hospital are specialist sickle cell centres in South London, offering a comprehensive range of care that includes: haematological, orthopaedics, renal care, health psychology, adolescents' transition, obstetrics, and neurology and pain management.

Sir Hugh Taylor, Chairman of Guy's and St. Thomas' Hospital (pictured left), took time out of his busy schedule to address the conference and to say a few words of welcome and good wishes to patients and their families.

The Patient Awareness Day at Guy's and St. Thomas' attracted over 74 patients, family members and friends and many colleagues attending throughout the day. The feedback from the patients was very positive stating that they were either satisfied or very satisfied with day.

Look out for the Patient Awareness Days in 2013! We hope to see you there!

RESEARCH UPDATE

Professor Swee Lay Thein

Consultant Haematologist, King's College Hospital and Professor of Molecular Haematology, King's College London

UPDATE ON CLINICAL STUDIES

Genetic modifiers of sickle cell disease

Chief Investigator – Prof. S L Thein

We are all only too aware of how differently sickle cell disease can manifest itself in different people and even within the same person at different periods of their lives. While environment and stress can trigger pain and constitute to some of the problems, we know that other co-inherited genes that influence the severity of disease. It is a challenge to be able to predict who is at risk of developing what complications so that we can try to prevent them BEFORE they happen.

We have already recruited more than 600 participants from King's College and Guy's and St. Thomas' Hospitals, BUT WE NEED MORE participants. We will also be recruiting patients in Lewisham and Queen Elizabeth Hospitals. **Please ask your clinician or clinical nurse specialist if you interested in taking part.**

Safety Study of MP4CO in Adult Sickle Cell Patients – Phase 1b (Sangart)

Chief Investigators – Prof. S L Thein & Dr Jo Howard

Sickle cell anaemia is caused by distortion of red blood cells but this can only take place under low oxygen conditions.

This study investigates if MP4CO, which is designed to carry small amounts of CO (a gas molecule), can have an oxygen therapeutic affect for reversing and terminating a sickle crisis.

The initial phase of the study will close in December 2012. Twelve patients in King's College Hospital and four patients in Guy's and St. Thomas' Hospital have taken part.

Vaccination against Pneumococcus in children

Chief Investigator – Dr David Rees

Paediatricians at King's College Hospital are involved in a trial of a new sort of immunisation against pneumococcal infection, with a vaccination which covers 13 different strains of the bacteria. This involved two vaccinations six months apart with blood tests to measure the response. This trial is organised by Pfizer who make the vaccine, and has nearly finished now, with no new patients being enrolled.

Study of determinants of severity of HbSC disease

Chief Investigator – Dr David Rees

Also at King's College Hospital, consultants are comparing different laboratory and clinical features of people with HbSC and HbSS disease, to try and understand more about both conditions, particularly HbSC. We are particularly looking at the way the red cell membrane works in these conditions. The study is being done jointly with Cambridge University, and is funded by the Medical Research Council.

A big thank you to you to all the patients who volunteered and taken part in these studies. If you would like to get involved, please ask your clinician next time you are in clinic.

Dr Jo Howard

Consultant Haematologist, Guy's and St. Thomas' Hospital

TAPS STUDY UPDATE

The Transfusion Alternatives Pre-operatively in Sickle Cell Disease study (TAPS) study ran from 2007 and 2011 and involved patients from all over the UK, Europe and Canada, including patients from Guy's and St Thomas' Hospital, King's College Hospital, University Hospital Croydon, University Hospital Lewisham and Queen Elizabeth Hospital Woolwich.

MANY THANKS TO ALL THOSE PATIENTS WHO PARTICIPATED IN THE STUDY.

It has helped us to answer several important questions about how best to manage patients with sickle cell disease (SCD) who are having surgery.

Before this trial it was not clear if patients should be offered blood transfusion before surgery. There did seem to be a high rate of complications, including sickle cell crisis, after surgery. In some reports, giving pre-operative transfusions before surgery was of benefit, but in other reports it was not.

This study looked at patients with HbSS and HbSβ^0 thalassaemia who were having low-risk or medium-risk surgery only. Medium risk surgery includes operations such as tonsillectomy, laparoscopic cholecystectomy and total hip replacement. Low risk surgery includes adenoidectomy, cataract surgery and hernia repair.

Patients were randomly divided into two groups, one group having pre-operative transfusion and the other group not having a transfusion. 67 patients completed the trial and it showed that patients who **did not** have a blood transfusion pre-operatively were more likely to have clinical complications during or after surgery. Clinically important complications were seen in 13 patients (39%) who did not have a pre-operative transfusion compared with only 5 patients (15%) who did have a transfusion. Most of these complications were acute chest syndrome, which is a life threatening complication of SCD. 12 patients who **did not** have a pre-operative transfusion required a blood transfusion during or after the operation compared with only three patients in the transfused group. Almost all the patients included in the stufy had HbSS rather than HbSβ^0 thalassaemia and over 81% of the operations were medium risk. Therefore it is difficult to draw conclusions about what we should do in patients with other types of SCD or who have low risk surgery.

In conclusion this study shows that rates of peri-operative complications are decreased by giving pre-operative transfusions in patients with homozygous sickle cell disease (HbSS) who are having medium risk surgery and pre-operative transfusions should be standard management in this patient group.

BRIDGING THE GAP: WORKING TOGETHER TO DEVELOP THERAPEUTIC EDUCATIONAL TOOLS FOR CHILDREN, YOUNG PEOPLE AND FAMILIES IN SOUTH LONDON

Dr Jo Blundell, Clinical Psychologist, Paediatric Sickle Cell & Thalassaemia Team, King's College Hospital

Four members of the South Thames Sickle Cell & Thalassaemia Network travelled to Paris in October to meet members of ROFSED, a team of multi-disciplinary professionals who are leading the way in the development of therapeutic educational tools aimed towards enhancing the lives and health care provision of children and young people with sickle cell disease in western France. The visit enabled us to share a range of innovative ideas about ways to engage and support children, young people and families in developing the knowledge and skills needed to manage their condition with confidence.

The provision of accessible and developmentally appropriate information is an important aspect of care for all children and young people with a physical health condition and plays an integral role in supporting a young person's transition to adulthood. As we know this period is a time of significant physical and psychological change for all young people, as they begin the process of developing autonomy, identity and social independence. For young people with sickle cell disease and/or thalassaemia, there may be additional stresses to manage including coping with unpredictable symptoms such as pain and fatigue, managing aspects of treatment and adjusting to school and work absences.

One of the greatest challenges for young people can be working towards achieving independence and autonomy in the management of their condition. To successfully make the transition to adult care, the health care team, young people and families need to work together as partners to ensure that young people develop the knowledge and skills needed to meet the challenges of managing their condition independently. Therapeutic Education Programs have been used to support children, young people and families with this process; enabling individuals to better understand their condition and facilitating the development of treatment specific knowledge and skills. It is recognised that the integration of therapeutic education alongside transition programs has measurable benefits for young people with a physical health condition in terms of improved management and treatment adherence, and enhanced quality of life (WHO, 1998).

This visit was the beginning of, what we hope will be, a productive and rewarding collaboration across services. The development of a multi-disciplinary working party focusing on taking forward the ideas developed from the visit was a positive first step, and we look forward to the coming year where we will continue to work together to enhance service provision for children, young people and families with sickle cell and/or thalassaemia living within South London.

We would like to thank the ROFSED team and clinicians from Necker Children's Hospital Paris for providing such an inspiring visit!

From left to right:
Sandra O'Driscoll, Sickle Cell Clinical Nurse Specialist, KCH; Jo Blundell, Clinical Psychologist, KCH; Jo Levitt CAMHS Practitioner, GSTT; Luhanga Musumadi, Sickle Cell Transition Nurse, GSTT.

Don't forget, you can email your comments about red cell *news* to: info@ststn.co.uk

HEALTH PSYCHOLOGY SERVICES

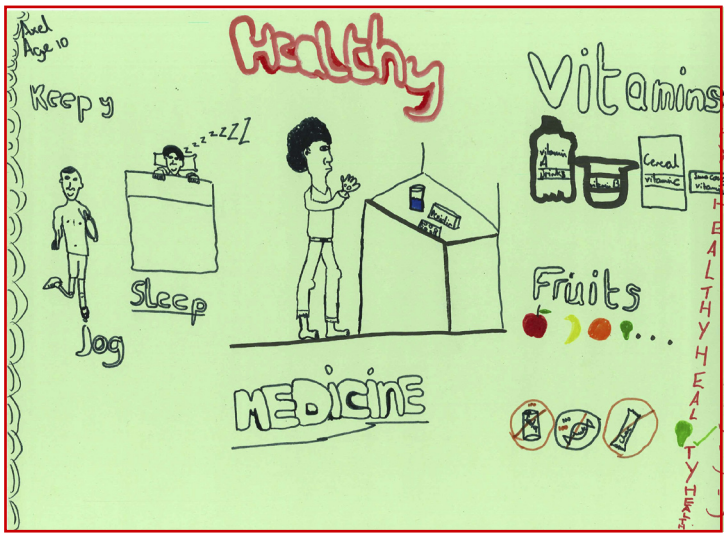
SICKLE CELL CHILDREN'S EDUCATIONAL GROUPS

Florence Bristow
Assistant Psychologist, Paediatric Sickle Team, Evelina Children's Hospital

The Paediatric Sickle Cell Team at the Evelina Children's Hospital host a series of Children's Educational Groups aged 10-14 with Sickle Cell Disease three times every academic year.

LOOKING AFTER OURSELVES

Our most recent group, held during the Autumn half-term had the theme "Looking After Ourselves' and the children drew pictures about what that' meant to them; identifying the medicines they take, the importance of exercise, healthy eating, keeping hydrated, getting rest and keeping warm.



We then played some games including 'the chocolate game' and a snakes & ladders quiz game which tested the children's understanding of their condition and how they can look after themselves to stay well. The games were great fun and an opportunity to facilitate discussion about the different techniques children use to remember to take their medication or how they might manage their condition at school, for example.

Dr. Inusa, Paediatric Consultant at the Evelina, also joined us for a group discussion where everyone could ask questions and share their ideas.

CHILDREN'S EDUCATIONAL GROUPS AIM TO:

- *Provide a platform for children to meet peers with the same condition*
- *Help children to learn about SCD*
- *Encourage children to think about managing their condition (as a precursor to preparing for transition from Paediatric to Adult services)*
- *Provide an opportunity for parents to meet and have a discussion together with members of the Sickle Cell Team*
- *Allow children and parents to ask questions that they might have otherwise found difficult to ask ('secret questions' are placed anonymously in a box and discussed together at the end of the afternoon*

Parents and children tell us that these afternoons are fun, enjoyable and valuable educational experiences. Parents added that it is beneficial for their child(ren) to meet other children with SCD and that the groups helped their child(ren) to learn more about SCD so that they don't feel that they are alone.

The next children's group will be held in the Spring half term holiday and we will send out information nearer the time. It would be great to see as many children as possible.

If you have any questions about the Evelina Sickle Children's Educational Group, please contact Florence: florence.bristow@gstt.nhs.uk or 0207 188 7774.

With thanks to our funders

King's College Hospital Charity · UK Forum on Haemoglobin Disorders · Roald Dahl's Marvellous Children's Charity – Making life better for seriously ill children · NOVARTIS