North East & Yorkshire Haemoglobinopathy Coordinating Centre

Sickle Cell Disorder

Patient Education Event

21st June 2025

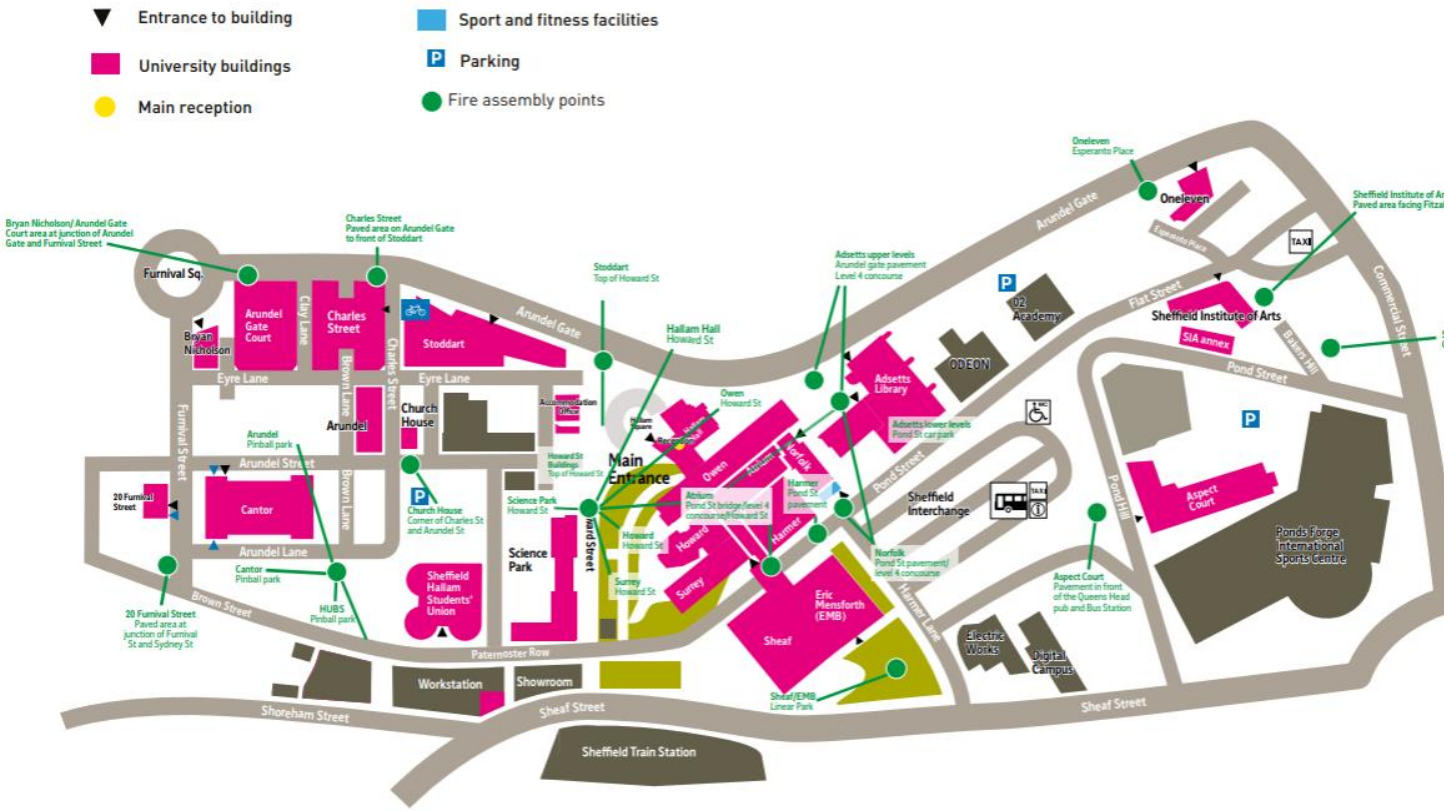
North East & Yorkshire Haemoglobinopathy Coordinating Centre

Welcome

Hallam Hall		
Time	Topic	Presenter
10:30 - 10:40	Welcome & Introduction	Dr Emma Astwood & Dr Clare Samuelson
10:40:11:00	Patient Rep Group	Blessing Olasolomon and Lizzy Okokwa Chair & Deputy Chair N E & Y HCC Patient Rep Group
11:00 - 11:30	Fertility and pregnancy	Dr Étienne Ciantar
11:30 - 11:50	Break - Rock choir Refreshments Chef Hallam Central	
11:50 - 12:30	Nutrition	Dr Claudine Matthews
12:30 - 13:00	New Treatments	Dr Clare Samuelson
13:00-13:10	Group Photo	
13:10 - 14:00	Lunch - Chef Hallam Central Visit stalls	
14:00-14:35	Mindfulness	Tony Watson
14:35-15:35	Blood donation docufilm / visit stalls	Hekima Asilia Thicker Than Water
	Refreshments available in Hallam Hall	
15:35-16:05	Closing Keynote Speaker	John James OBE Sickle Cell Society
16:05-16:15	Closing remarks	Dr Emma Astwood & Dr Clare Samuelson

Housekeeping

North East & Yorkshire Haemoglobinopathy Coordinating Centre



FIRE PROCEDURES

1 A **CONTINUOUS** siren means **EVACUATE** the building

1.1 leave the building quickly and quietly by the nearest available exit route.

*Wheelchair users should use move to the refuge point in the lift lobby area on level 6.

1.2 follow instructions given by the security staff or fire marshals immediately

1.3 the muster point is at the top of Howard Street.

at the muster point there will be a Fire Marshall, identifiable by wearing a high-vis fire marshall vest

1.4 do not re-enter the building until informed to do so

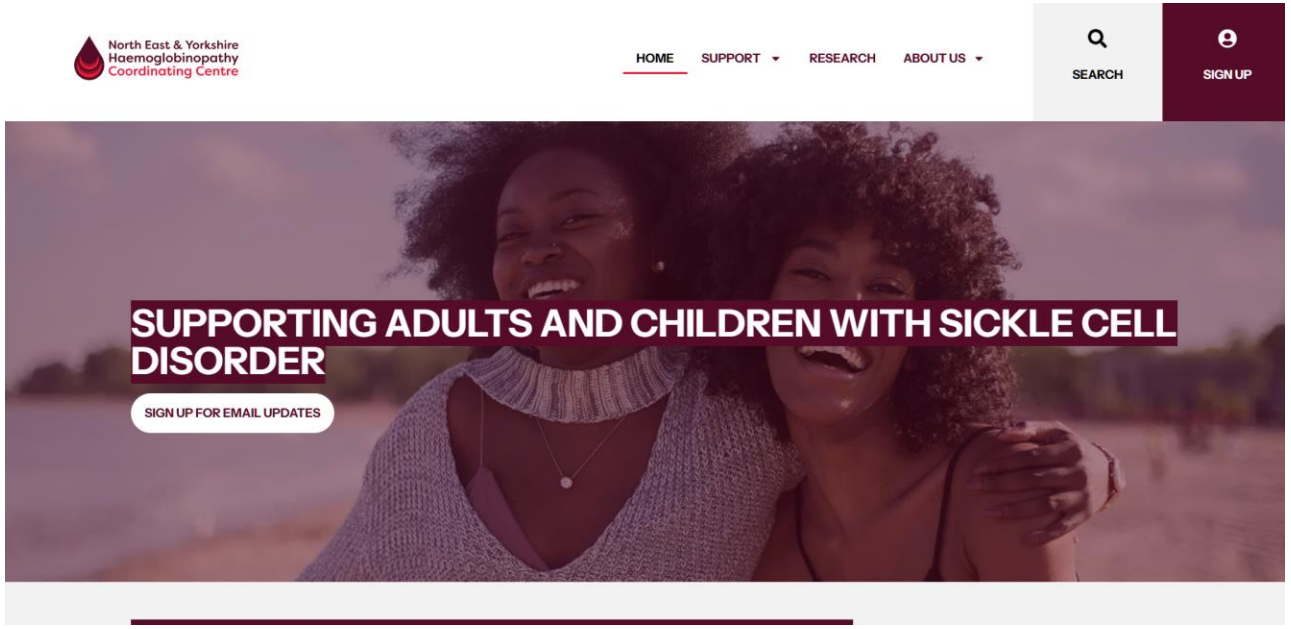
DO NOT use the lifts

DO NOT GO TO Level 10 – Children will be brought by staff.

Website

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Refreshed website
- Photos
- Speakers will reference it today
- QR codes on tables
- Sign Up!



North East & Yorkshire Haemoglobinopathy Coordinating Centre

NE&Y HCC Network Patient Representatives

Purpose of NE&Y HCC Network Patient Representative

North East & Yorkshire Haemoglobinopathy Coordinating Centre

The purpose of this role is to ensure that the Northeast & Yorkshire (NE&Y) Haemoglobinopathy Coordinating Centre (HCC) network actively listens to and learns from the experiences of individuals living with Sickle Cell Disorder (SCD) and their carers.

The Patient Representative Chair, Deputies, and the wider patient representative panel, with the support of the NE&Y HCC Network, facilitate regular meetings to gather feedback, share positive ideas, and provide constructive input to help shape and improve the services delivered by the network.

The group is made up of individuals with direct lived experience of Sickle Cell Disorder (SCD), whether through managing the condition personally or supporting a family member or friend affected by SCD or sickle cell trait. Our insights play a vital role in shaping patient-centered care and service improvement.

Main Duties

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Develop effective methods and forums to gather the views and experiences of SCD patients and carers across the NE&Y HCC region, ensuring these insights inform the NE&Y HCC work plan.
- Review SCD-related guidance regularly and evaluate its impact on patient and carer outcomes.
- Identify and share best practices to promote high-quality care throughout the network.

- Escalate immediate concerns or risks to the NE&Y HCC Network Manager for prompt resolution.

- Provide ongoing feedback to the NE&Y HCC Business Meetings, highlighting key areas that require further attention and action. Etc..

Overview of the Patient and Carer Representative Team

North East & Yorkshire Haemoglobinopathy Coordinating Centre

The group consists of 28 members in total, including 1 Chair, 3 Deputies, 23 patient and carer representatives' members, and 1 multidisciplinary team (MDT) representative who also serves as the Data Manager and Engagement Coordinator. All roles within the group are undertaken on a voluntary basis

Patient Representatives

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Chair: Blessing Olasolomon is a global student recruitment strategist and a passionate advocate for Sickle Cell Warriors. With an MSc in Advertising and Marketing, she blends educational expertise with lived experience to create meaningful impact. As Chair of the NE&Y HCC Patient Voice, Blessing is committed to empowering underrepresented voices, and championing health equity worldwide.

Dept Chair: Christobel Iwoba, is a dedicated Healthcare Assistant and Support Worker committed to sickle cell advocacy and patient well-being. She aims to become a haematology or mental health nurse, driven by a purposeful approach to life and work.

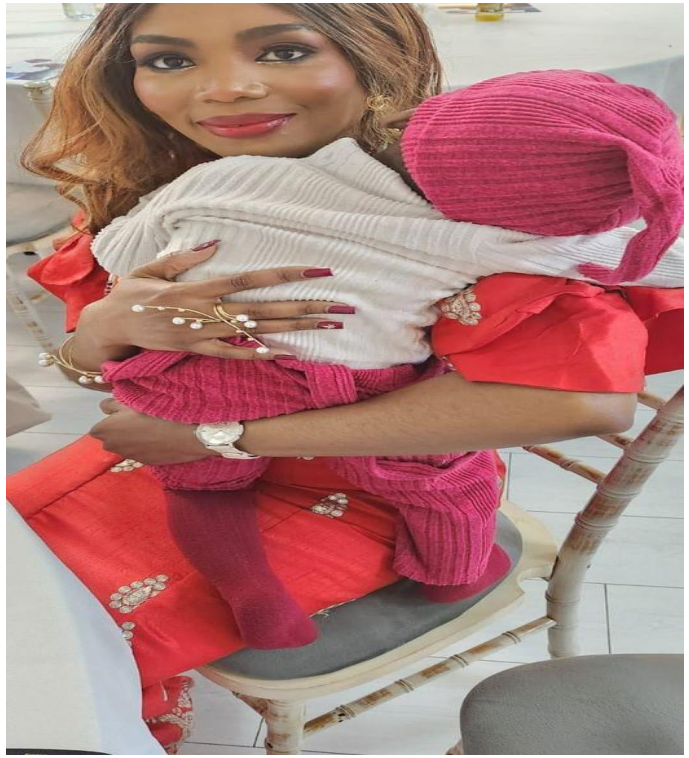
Dept. Chair: Elizabeth Okokwa, is an expert with extensive experience in global healthcare initiatives. Elizabeth has collaborated with international medical organizations focusing on disease prevention and community healthcare. Now a Charity Administrator in Yorkshire, Elizabeth's expertise, experience and her personal journey with her young daughter, a resilient sickle cell warrior, drive her dedication to advancing sickle cell care and advocating for equitable health solutions.

Dept. Chair: Johnson Ochai, is a Solicitor with Liberty Solicitors. He brings a strong legal and advocacy background to his role as Deputy Chair for Patient Representation at NE&Y HCC. His focus is on ensuring that every voice within the sickle cell community is valued.

Blessing's background and motivation

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Diagnosed with Sickle Cell Anaemia at age 7 in Nigeria, my journey has been marked by resilience and hope. Since moving to the UK in 2022, I have faced numerous challenges , from chronic crisis pain and multiple ICU stays to chronic leg ulcers and a traumatic pregnancy in 2023.



Managing sickle cell is not just about surviving , it's about living fully, with dignity, support, and a powerful voice. That's why I joined and serve as Chair of the Patient Public Voice. No one living with sickle cell should ever feel invisible.

For me, representation isn't just important, it's life-changing.

In 2025, I experienced a stroke scare, a moment of profound fear not just for myself, but for my daughter. Today, I undergo monthly red cell exchange transfusions to reduce that risk. It's exhausting, but it fuels my determination.

Christobel’s background and motivation

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Daughter, sister, friend, Healthcare Support Worker, and Deputy Chair for Patient Representation at NE&Y HCC.

Diagnosed with sickle cell anemia at birth, I’ve faced chronic pain, multiple hospital stays, blood transfusions, and thigh ulcers. A portacath helps ease treatment challenges. Living with sickle cell is a constant physical, mental and emotional battle, often made harder by not being believed.

I’m committed to advocating for compassionate, patient-centered care. Through my lived experience, I strive to ensure everyone with a haemoglobin disorder is heard, respected, and valued ,in the Northeast & Yorkshire and beyond

Lizzy's background and motivation

North East & Yorkshire Haemoglobinopathy Coordinating Centre

I'm a mother to a vibrant, courageous young daughter living with Sickle Cell Disorder. She was diagnosed at the age of 3 years, and since then, the journey has been both challenging and transformative.

Balancing full-time work, caring for three other children, and managing the many demands of daily life can be overwhelming , but her care and well-being remain my absolute priority. Like many parents and carers, I often find myself stretched thin , emotionally, physically, and mentally. But I show up every day because my daughter deserves not just to survive, but to thrive. She deserves a future filled with opportunity, dignity, and compassionate care.

I joined the Patient Public Voice group because representation matters. Every family affected by sickle cell deserves to be heard and to help shape the care they receive. I'm here to be that voice, not just for my daughter, but for every family on this journey.

Johnson’s background and motivation

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Lived with sickle cell all my life, my motivation isn’t to retell what many already know, it’s to help build better systems, stronger patient engagement, and a culture of listening within healthcare.
This role gives me the opportunity to represent the interests of patients across the region, inspire confidence in others living with haemoglobin disorders, and contribute to positive, patient-led change. My focus is on collaboration, compassion, and making sure that lived experience is valued as expertise in its own right.

NE&Y HCC (PPV)– **Our Impact and Actions**

North East & Yorkshire Haemoglobinopathy Coordinating Centre

As Patient Public Voice (PPV) representatives, we are actively involved in shaping better care for the sickle cell community. So far, we've had the privilege to:

- Contributed practical ideas for *Winter Care Packs* to support Sickle Cell patients in the NE&Y region.
- Helped shape better patient communications, including contributing to the development of a new, more accessibl website.
- Shared lived experiences at NHS patient education events to promote better understanding and patient-centered care.

NE&Y HCC –Involvement so far

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Present at NHS Staff Education Days, bringing the patient voice directly to healthcare professionals
:
- Raise important concerns about emergency care, chronic leg ulcers, and the ongoing stigma around pain.

We engage. We speak up. We improve- together.
And we're just getting started.

Stay connected:
🌐 www.ney-hcc.co.uk/patient-engagement

Why Your Voice Matters

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Medical expertise saves lives , but *lived experience transforms care*.
As patients and carers, we *know* what it feels like:

:

- The unbearable pain of a crisis that words can't describe.
- Navigating endless A&E visits, trying to explain fatigue that never truly goes away.
- Facing stigma, isolation, mental health struggles, and strained relationships.
- Missing out on life simply because hospital visits become the norm.

But your voice can change that.
When we show up, speak up, and share our experiences, systems listen, and care improves.

Ready to be a part of Positive Change?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Your Voice Matters - Join Us

We need more voices. More stories. More strength. Whether you're living with sickle cell, a parent, or a carer, *your experience can help drive change*.

Be part of the movement.
"Together, we can shape a better, fairer future for everyone affected by sickle cell"

Join us today:
https://www.ney-hcc.co.uk/patient-public-voice-group-ppv-application-form/

Real voices. Real change.

North East & Yorkshire
Haemoglobinopathy
Coordinating Centre

Dr Étienne Ciantar MD, FRCOG, FHEA
Consultant – Obstetrics, Obstetric Haematology & Medical Education
Leeds Teaching Hospitals NHS Trust

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Fertility and Pregnancy

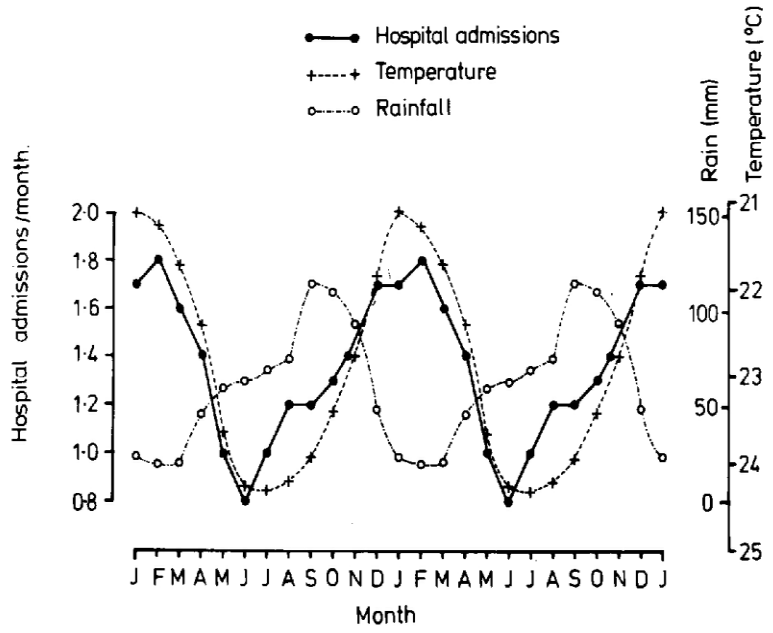
PATIENTS AND HOSPITALS

“for sickle cell patients, positive experiences of hospital care are the exception”

- mistrust
- control
- stigmatisation
- neglect

Precipitants of Sickle Cell Pain

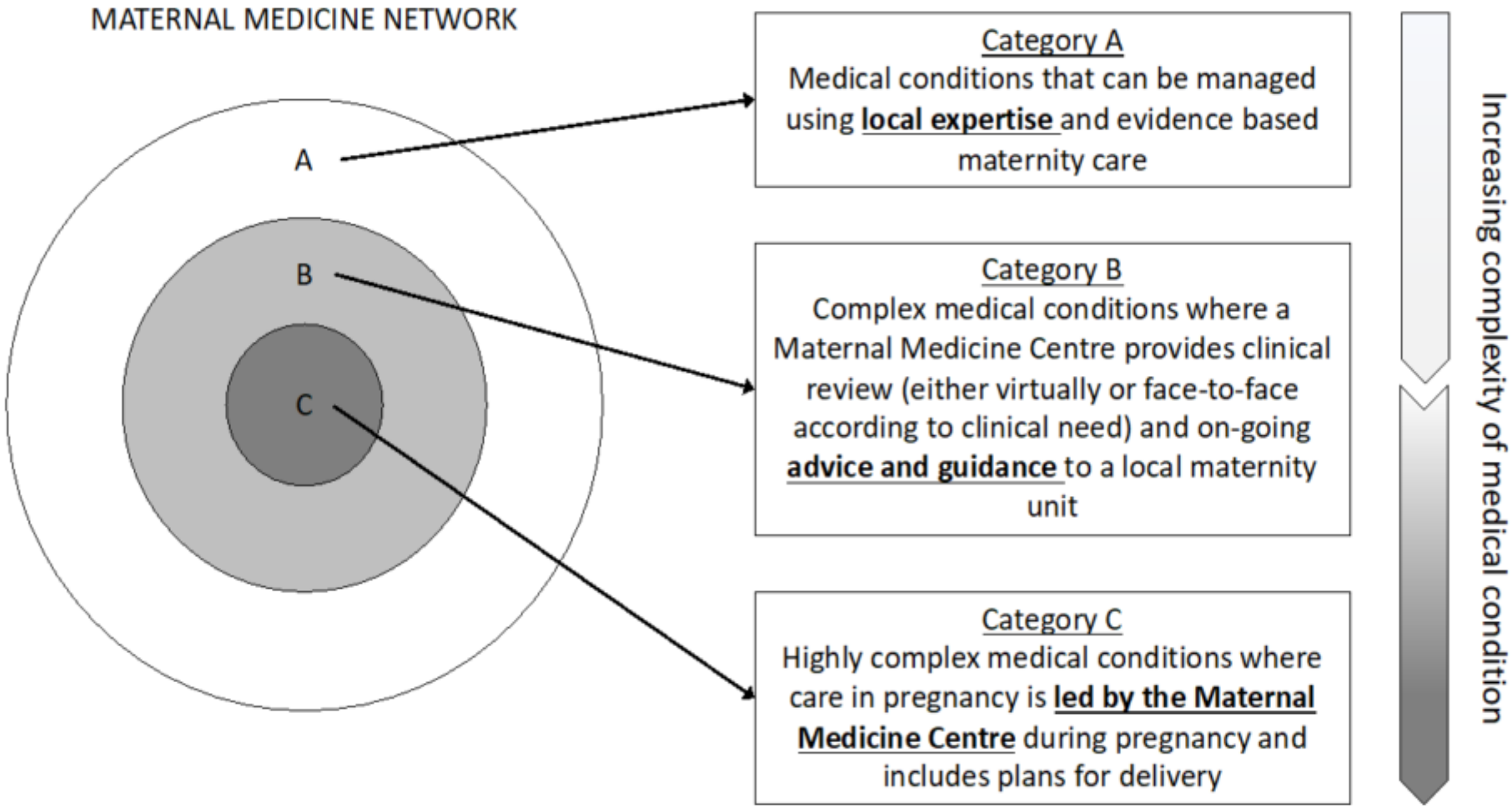
- Dehydration
- Infection
- Climatic
- Psychological
- Pregnancy



High risk situations

- Any patient with painful crisis
- Post operative
  - abdominal surgery
- Pregnancy
  - final trimester & post-partum

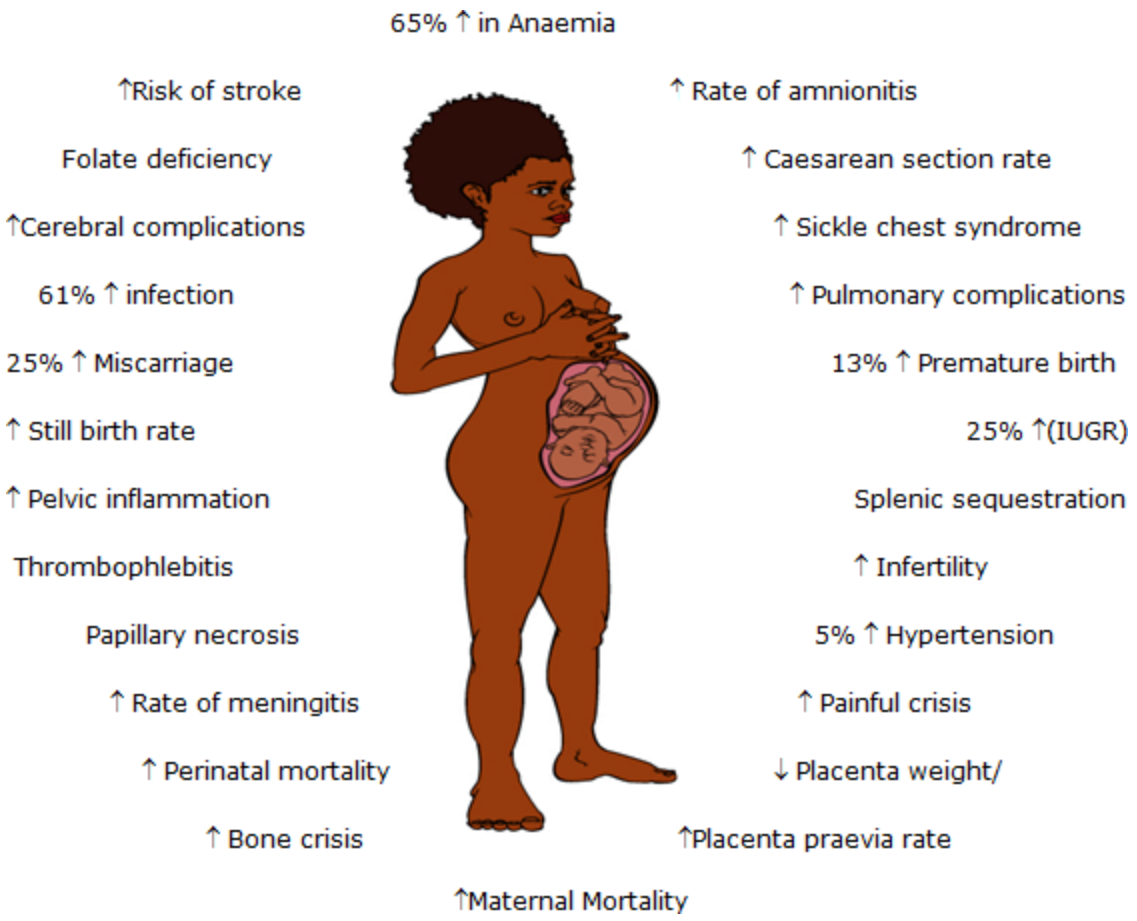
Obstetrics
Haematology
Specialist Midwife
Specialist Nurse
Anaesthetist
Neonatology
Blood Bank



Maternal Medicine Network

Haematological disease		
Sickle cell trait	Current immune thrombocytopenia and platelet count ≤75	Sickle cell disease
Historical immune thrombocytopenia and platelet count >75	Thrombocytosis	Beta thalassaemia major
Gestational thrombocytopenia	White cell disorders	Other complex thalassaemia: • iron overload • endocrine disease • pulmonary hypertension*
Current VTE or previous single VTE	Recurrent VTE	Current extensive VTE without other access to Factor Xa monitoring
Obstetric antiphospholipid syndrome	Thrombotic antiphospholipid syndrome	Antiphospholipid syndrome with extensive arterial events
Inherited thrombophilia (no VTE, not antithrombin deficiency)	Inherited thrombophilia with previous VTE	Antithrombin deficiency
History of treated haematological malignancy	Stable myeloproliferative/myelodysplastic disease	Active haematological malignancy
Alpha/beta thalassaemia trait	Mild, isolated clotting factor deficiency • Factor II, V, XI or XIII > 0.2iu/ml • Factor X > 0.3iu/ml	Clotting factor deficiency: • Factor II, V, XI or XIII ≤ 0.2iu/ml • Factor X ≤ 0.3iu/ml • Combined deficiencies
B12/folate deficiency	Mild platelet function disorder with platelet count >100	Moderate/severe platelet function disorder or with platelet count >100
	Carriers of haemophilia with known female fetus and normal factor VIII/IX	Carriers of haemophilia with male or unknown gender of fetus
	Type I Von-Willebrand disease, VWF activity normalised in pregnancy	Von-Willebrand disease: Type 1 if VWF not normalised, Type II and Type III

Sickle cell and pregnancy



Maternal morbidity

- Increased risk of
    - Hypertension and pre-eclampsia
    - Acute painful crisis: 20-56%
    - Anaemia
    - Infections (especially UTI: 16-23%)
    - Acute chest syndrome (11-17%)
    - VTE (Increase in DVT not PE – US cohort)
    - Caesarean section: 30-62%

Increased maternal mortality

- Range from 0% to 9.2% in studies of centres across the world
  - UK national data: approx. 1 death per year
    - UKOSS study 2010-2011 no deaths in 108 pregnancies
    - However, 3 reported deaths in 2016
  - US Co-operative study (1980's, 1990's)
    - 0.4% mortality
  - US In-patient sample 2000-2003
    - 72.4 deaths per 100,000 in SCD (0.07%)
    - 12.7 deaths per 100,000 in all pregnancies(0.01%)

Fetal complications

	Perinatal mortality	Fetal growth restriction	Prematurity
Ngo: SS	2.1%	14%	16%
Yu	1.4%	18%	24%
Chase	2.4%	20%	19%
UKOSS	2.7%	-	-

HbSS vs HbSC

- HbSC tend to have milder phenotype
  - Jamaica (95 pregnancies)
    - No difference in pre-eclampsia, stillbirth rate, mean birthweight from AA controls
    - Sickle related complications similar to SS
  - France (33 pregnancies)
    - No difference in obstetric outcomes and perinatal outcomes from AA
    - Pain in 36% (cf 42% of SS)

- Decreased risk of maternal and fetal complications in HbSC

BUT

- Although most are mildly affected, some do have serious complications
- Not possible to predict who will have severe complications

Therefore treat in same way as HbSS

Can intervention improve outcome?

- Benin, West Africa
- June 1993-Dec 1993
  - 15 pregnancies
  - 4 maternal and fetal deaths/2 fetal deaths (27%/40%)
- Feb 1994 – Dec 1997. Prospective study
  - Education, nutrition, malaria prophylaxis and antibiotics
  - 108 pregnancies
  - 2 maternal deaths (1.8%), 13 fetal deaths (11.9%)

Rahimy et al 2000

bjh guideline

Management of sickle cell disease in pregnancy. A British Society for Haematology Guideline

Eugene Oteng-Ntim,[1,2] Sue Pavord,[3] Richard Howard,[4] Susan Robinson,[1] Laura Oakley,[5,6] Lucy Mackillop,[3] Shivan Pancham,[7] Jo Howard[8,9] on behalf of the British Society for Haematology Guidelines Committee

[1]Department of Women's Health, Guy's and St Thomas' NHS Foundation Trust, [2]Department of Women's Health, King's College London, [3]Department of Haematology, Oxford University Hospitals NHS Foundation Trust, Oxford, [4]Department of Obstetrics and Gynaecology, Barking, Havering and Redbridge University Hospitals, Romford, [5]London School of Hygiene and Tropical Medicine, London, [6]Centre for Fertility and Health, Norwegian Institute of Public Health, Oslo, Norway, [7]Department of Haematology, Sandwell and West, Birmingham Hospitals NHS Trust, London, United Kingdom of Great Britain and Northern Ireland, [8]Department of Haematology, Guy's and St Thomas' NHS Foundation Trust, and [9]Department of Haematology, King's College London

Keywords: sickle cell anaemia, sickle cell disease, pregnancy, antenatal, intrapartum, preconceptual.

This guideline was compiled according to the British Society of Haematology (BSH) process at https://b-s-h.org.uk/guidelines/proposing-and-writing-a-new-bsh-guideline/. The Grading of Recommendations Assessment, Development and Evaluation (GRADE) nomenclature was used to evaluate levels of evidence and to assess the strength of recommendations. The GRADE criteria can be found at http://www.gradeworkinggroup.org

Literature review details

This BSH guideline was developed and updated from a previous Royal College of Obstetricians and Gynaecologists (RCOG) Green-top guideline[1] in accordance with the standard method of producing BSH guidelines. Medline, Embase, the Cochrane Database of Systematic Reviews, the Cochrane limited to humans and the English language. The National Library for Health and the National Guidelines Clearing House were also searched for relevant guidelines.

Review of the manuscript

Review of the manuscript was performed by the BSH Guidelines Committee, General Haematology Task Force, the BSH Guidelines Committee and the members of the sounding board of BSH. It was also placed on the members' section of the BSH website for comment. It has also been reviewed by the Royal College of Obstetricians and Gynaecologists, Sickle Cell Society and BSH Obstetric Haematology Special Interest Group; these organisations do not necessarily approve or endorse the contents.

SCD and pregnancy

Pre-conception

Counselling +/- partner testing

Contraceptive advice

- **1st choice: progesterone only (pill, depot, implant)**
- **2nd choice: copper IUD, combined pill**

Fertility – sperm count/mobility, impotence

Medications – hydroxycarbamide, ACE inhibitors

Screen for red cell allo-antibodies & iron overload (if transfused), review vaccination status, record full red cell phenotype, consider ECHO if not done in last year.

Pregnancy complications

Maternal

Increased incidence of:

- Infection (UTI, pneumonia, puerperal sepsis)
- Painful crisis
- Acute chest syndrome (Fever, tachypnoea, pleuritic chest pain)
- Venous thromboembolism
- Preterm labour
- Pre-eclampsia
- Caesarean section
- Maternal death (0-9%, UKOSS 0%)

Fetal

Increased incidence of:

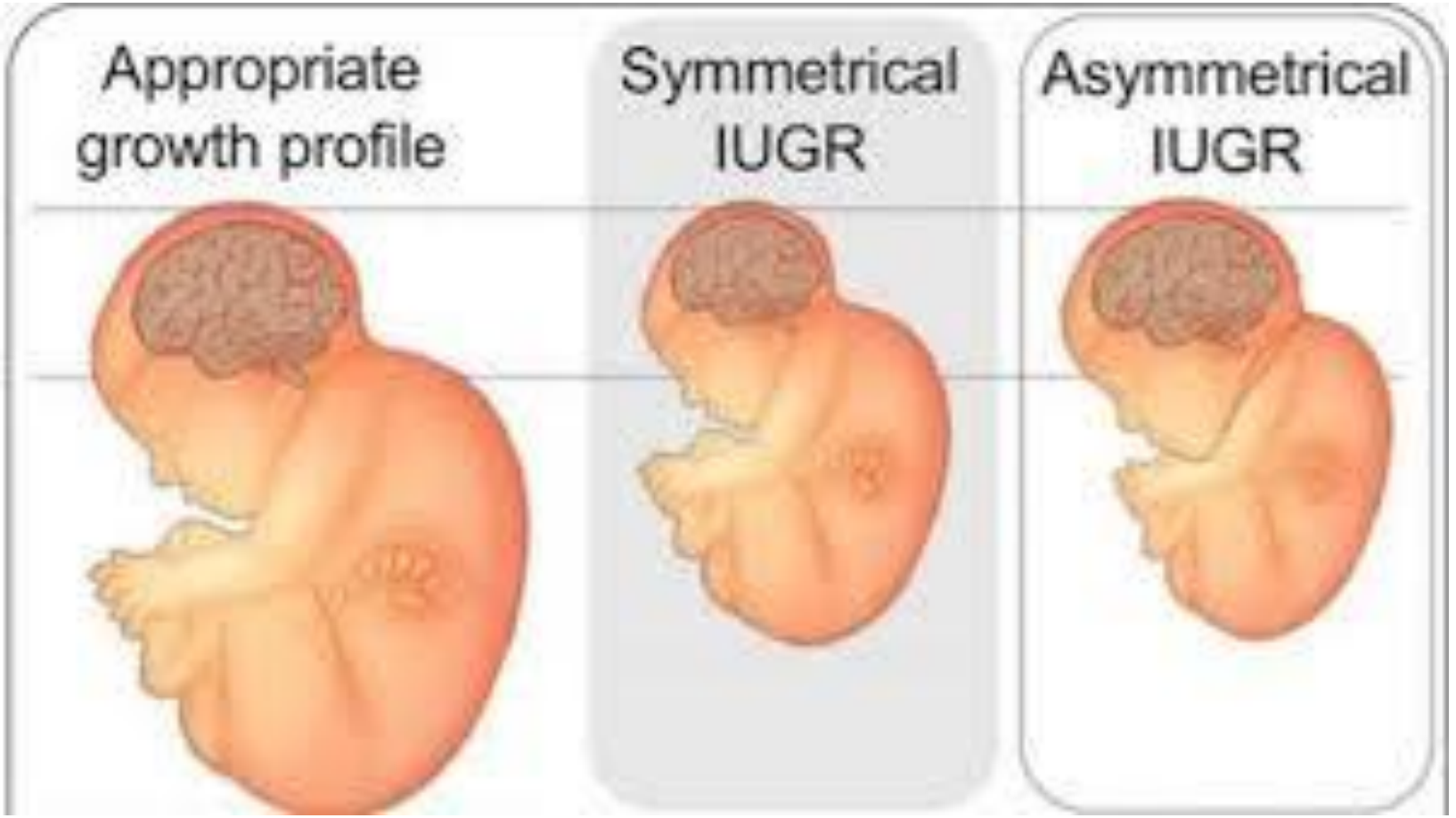
- Miscarriage
- Intra-uterine growth restriction
- Prematurity
- Fetal distress
- Stillbirth/neonatal death (perinatal mortality 15% ie: 15x general population)

Antenatal review

- Clinic schedule in a specialised tertiary unit setting
  - 4 weekly to 24 weeks
  - 2 weekly to 38 weeks (alternating midwife and obstetric team)
  - Then weekly
- Ensure on folic acid 5mg and penicillin prophylaxis
- LMWH blood thinning injections from 28 weeks or before
- Aspirin 150mg from 12 to 36 weeks
- Oral iron if lab evidence of deficiency

Antenatal monitoring

- Ultrasound:
  - Early viability scan at 7-9 weeks gestation
  - Routine first-trimester dating scan at 11-14 weeks +/- Downs screening
  - Anatomy scan at 20 weeks
  - Four weekly scans for fetal growth, liquor volume and placental function from 24 weeks


Appropriate growth profile
Symmetrical IUGR
Asymmetrical IUGR

Antenatal monitoring

Check	Check FBC and MSU monthly (blood pressure and urine dipstick at each visit)
Discuss	Discuss suitable positions for delivery if patient with a hip replacement
Notify	Notify neonatologist in writing if neonatal complications expected
Consider	Consider induction of labour at 38-40 weeks

Blood transfusion

- Prophylactic blood transfusion remains controversial
- TAPS-2 study – serial prophylactic EBT may lead to less VOC, preterm labour and improved baby birth weight

- *<u>Possible indications</u>:*

- Recurrent crisis, Hb <70g/l, previous poor obstetric history, patients on hydroxycarbamide pre-conception, multiple pregnancy

- HbSS more likely to require transfusion
- All transfusions should be discussed with the haematology team
- Transfused blood should be matched for Rh, Kell and be Hb S and CMV negative.

Labour and delivery

- Prolonged labour (>12 hours) should be avoided, with early recourse to caesarean section as increased stress may trigger a crisis
- If painful crisis occurs, give morphine
- Continuous CTG
- Choice of analgesia: patient's choice, but to minimise distress, have a low threshold for epidural
- PCAS may be considered
- Advise neonatologist of delivery

Postnatal care

- Keep warm and well hydrated
- Chest X-ray if any chest pain or low saturation, because of risk of sickle chest
- Manage any infections quickly
- Good pain control (e.g. morphine, PCA)
- Anticipate the need for laxatives and anti-emetics
- LMWH blood thinning injections for 6 weeks
- Ensure good follow up (community midwife and haematology clinic)

Conclusions

- SCD is a high-risk condition in pregnancy
- Care needs to be provided in a specialist centre
- MDT approach
- Guideline driven care but with an individual patient-centred approach
- 300 babies are born each year in the UK from SCD mothers

Your Diet Your Health:

Healthy Eating Principles to improve Your Health Outcomes

Presented by

Dr Claudine Matthews DPROF HSC RD FHEA

NE & Y Patient Education Event

21st June 2025

Education Session (1) Agenda:

Part 1: How sickle cell affects your nutrition

Part 2: Why is good nutrition important

Part 3: Improving your nutritional intake using the Sickle Cell Nutrition Eatwell guide

Part 4: Benefit of Good nutrition

My Background

- South Africa
- Apartheid – experienced racism, marginalisation and disempowerment
- Worldview of social justice, advocacy and empowerment
- Moved to the UK in 2000
- Introduced to sickle cell in 2011 and sickle cell nutrition in 2014
- Started my doctorate in 2016 and completed in 2023
- Founded – Sickle Cell Nutrition Academy in 2023
- Consultant Dietitian –
  - Leading education and guidance, advising on complex nutritional problems, research and innovation - advancing sickle cell nutrition

Part 1: How sickle cell affects your nutrition

Rationale for Sickle Cell Nutrition?

- Cause
  - Ongoing break down of your red blood cells
  - Sickle cell crisis causes tissue and organ damage
  - Increased risk of infection
- Impact
  - Chronic anaemia/fatigue
  - Multiple Nutrient deficiencies
  - Poor nutritional
  - Impaired immunity
- Management
  - Eating a Healthy Balanced Diet
  - Engage in healthy lifestyle activities like regular activity
  - Don’t smoke, reduce alcohol intake

Table One: Main Clinical Manifestation of affected Organs and Systems[2]

System/Organ	Clinical Manifestation/Complication
Skeletal	Osteonecrosis, Osteomyelitis, leg ulcers, avascular necrosis
Genitourinary	Chronic renal insufficiency, priapism, chronic renal failure
Gastrointestinal	Cholelithiasis, viral hepatitis from transfusion, liver failure
Spleen	Splenic enlargement and fibrosis, acute aplastic anaemia, parvovirus B19, functional asplenia, leucocytosis
Hepatobiliary	Indirect hyperbilirubinemia
Caridiopulmonary	Pulmonary hypertension, cardiomegaly, cardiac failure
Central nervous	Stroke, silent infarcts

Your sickle cell complications can affect your Nutritional Needs

Common Nutritional Problems

Common Nutritional Problems

1. Malnutrition
2. Aneamia/Fatigue
3. Impaired Immunity/Chronic Inflammation
4. Dehydration/ Constipation
5. Iron Overload
6. Osteoporosis
7. Increased risk of cardiovascular disease

Part 2: Why is good nutrition important?

Why is good nutrition important?

Building a good Nutritional Status and preventing malnutrition:

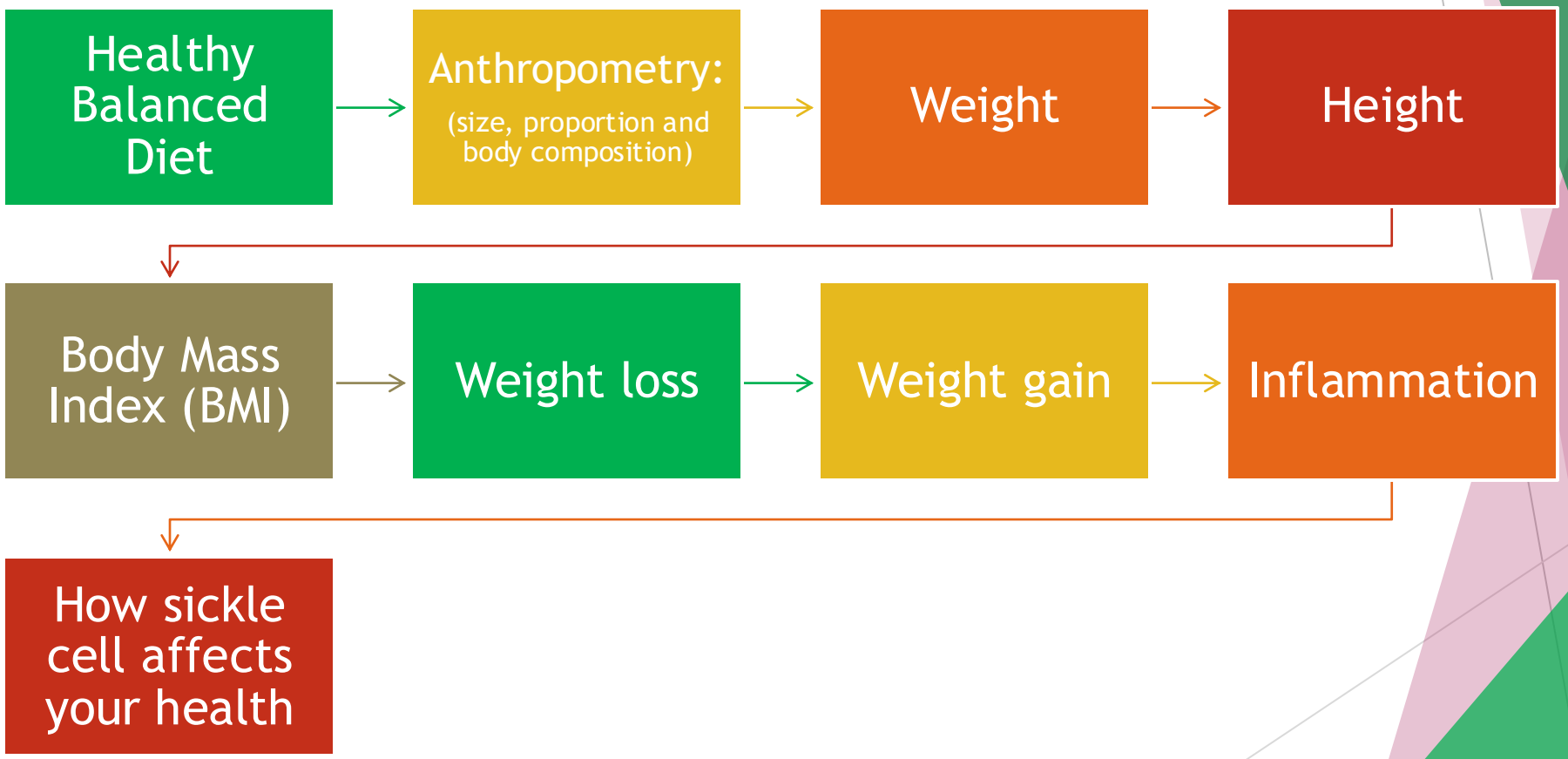
1. **Eating a balanced** diet:
   - Eat the right types of food
   - Eat right amount of food
   - Eat the right variety of food

2. Choose food from all the food groups:
   - Ensure the right balance of nutrients to meet your bodies needs

3. Help your body to function as **Optimally** as possible

4. Enjoy a **good quality of LIFE** and do the things that you **Enjoy doing.**

What is needed for a good nutritional Status



Healthy Life Expectancy

- **Blue zone principles (Dan Buettner)**
- 9 principles:
- 1st…Move naturally
- 2nd…Have a purpose
- 3rd …Downshift
- Find out the other 6 and tell me to win a surprise prize…

Part 3: Improving your nutritional intake using The Sickle Cell Nutrition Eatwell Guide

SICKLE CELL NUTRITION EATWELL GUIDE

Tailoring diet and nutrition knowledge and information to Sickle Cell Disease by increasing understanding of cultural and more sustainable foods enabling patients to better balance their diet and enhance health outcomes

CHOOSE HEALTHY OPTIONS

Use the traffic light system

Choose foods lower in fat, salt and sugars

Eat at least 5 portions of a variety of fruit and vegetables every day

Fruit and vegetables

TAMARIND

PAK CHOI

SOURSOP

OKRA

Regulate Immunity– include anti-oxidant foods

Reduce Inflammation

Choose a wholegrain or higher fibre versions with less added fat, salt and sugar

Potatoes, bread, rice, pasta and other starchy carbohydrates

BREAD

HARD DOUGH AND AGEGE

TARO ROOT

GREEN BANANA

POTATOES / SWEET POTATOES

YAMS

PORRIDGE

PLANTAIN

Include Energy Boosting Foods

PASTA

RICE

FUFU BANKU/ GARRI KENKEY

ROTI, WAJERA

WHOLEGRAIN FLOUR

CORNMEAL

CHICKEN

CHICKPEAS

LENTILS

BEANS

Increase Protein Intake

MEAT (LEAN MINCE OR)

FISH

EGGS

Beans, pulses, fish, eggs, meat and other proteins

Eat more beans and pulses, 2 portions of sustainably sourced fish per week, one of which is oily. Eat less red and processed meat

LOW FAT MILK

Improve Bone Health

Dairy and alternatives

Choose lower fat and lower sugar options

Eat less often and in small amounts

MALT BEVERAGE

PIES, QUICHE

MEAT PIES

FRIED CHICKEN

CHOCOLATES AND SWEETS

PREVENT DEHYDRATION

8-10 per day

Water, lower fat milk, sugar-free drinks including tea and coffee all count.

Limit fruit juice and/ or smoothies to a total of 150ml a day.

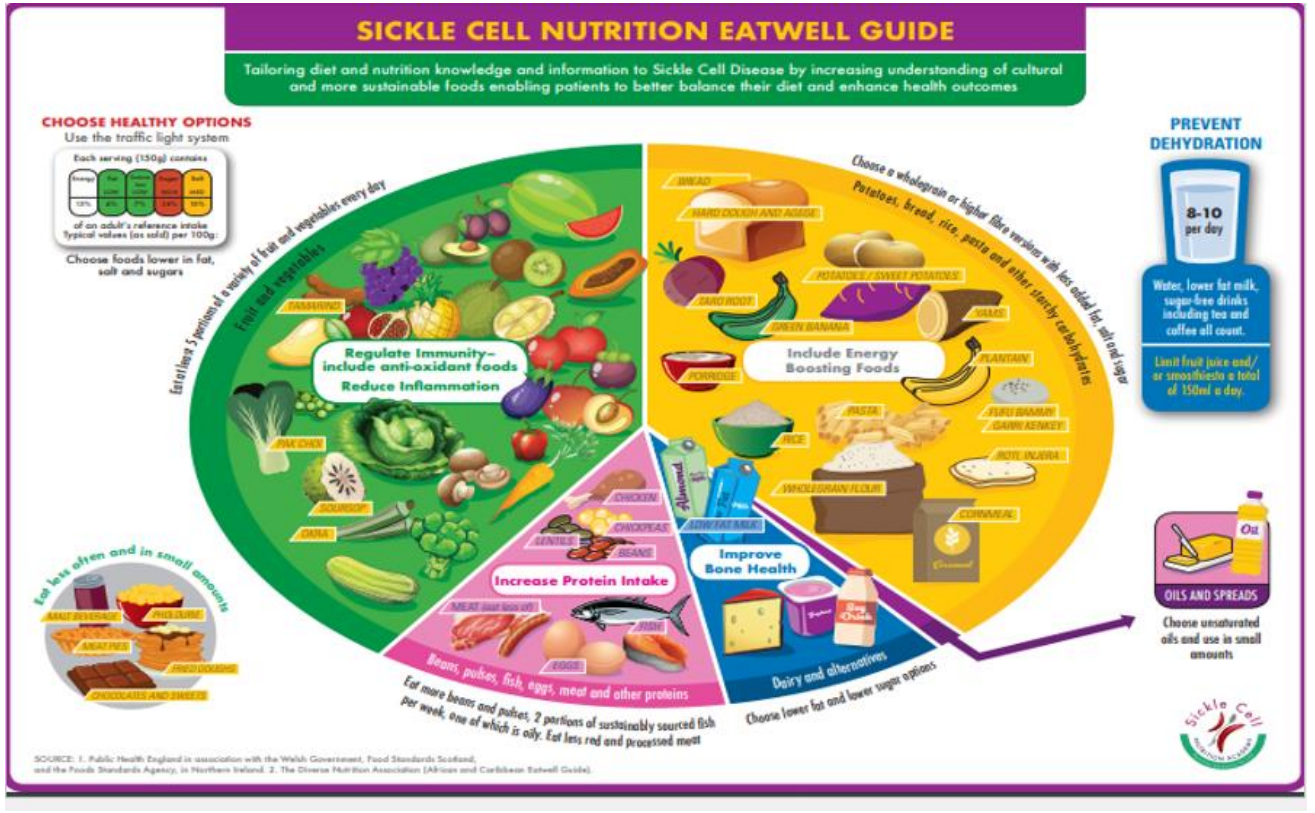
OILS AND SPREADS

Choose unsaturated oils and use in small amounts

SOURCE: 1. Public Health England in association with the Welsh Government, Food Standards Scotland, and the Foods Standards Agency, in Northern Ireland. 2. The Diverse Nutrition Association (African and Caribbean Eatwell Guide).

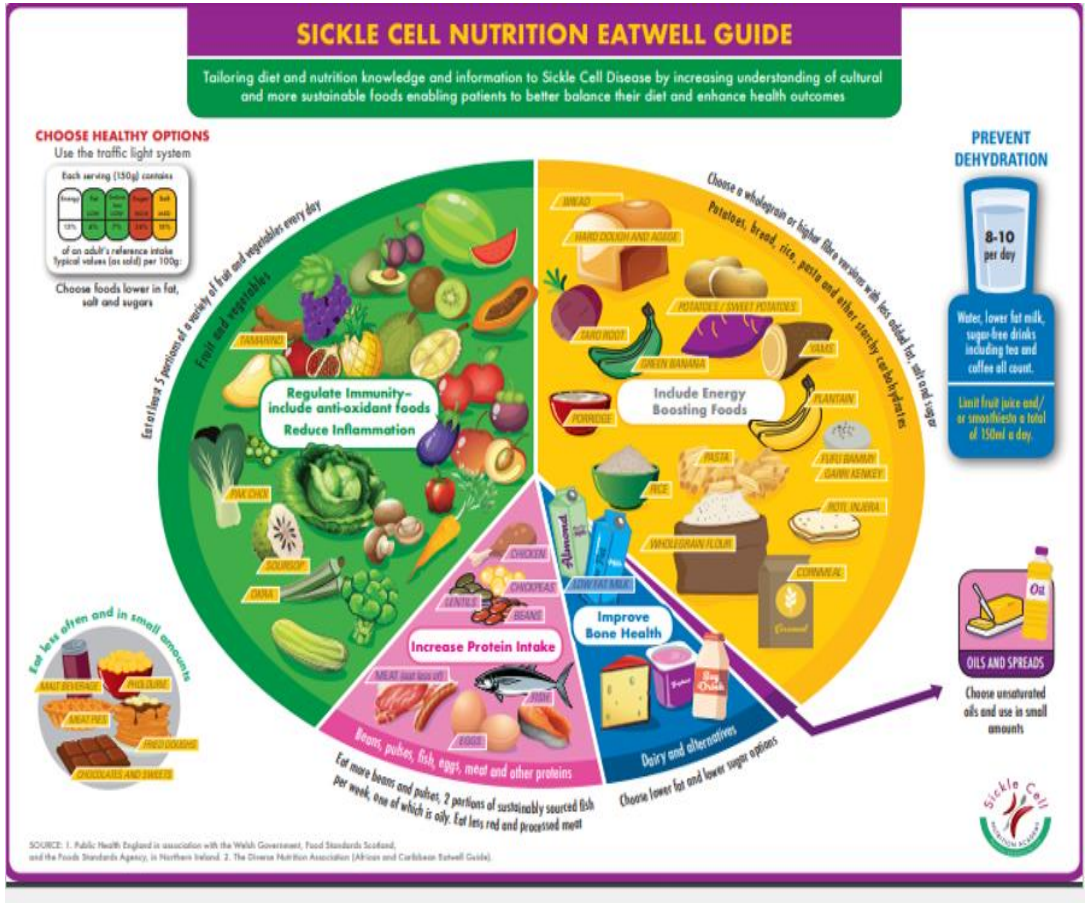
7 Key Healthy Messages in the SCD Eatwell Guide

- 1. Include energy boosting foods
- 2. Increase protein intake
- 3. Regulate immunity -include antioxidant foods
- 4. Reduce Inflammation
- 5. Improve bone health
- 6. Prevent Dehydration
- 7. Choose Healthy options



5 Food Groups

- Carbohydrates (CHO)- breads, cereals, potatoes, rice, pasta, yam, plantain
- Fruit and Vegetables (Your 5 a Day)
- Proteins - Beans, pulses, meats and other protein, fish, eggs, other proteins
- Dairy, milk and milk alternatives, cheese, yogurt
- Fats and oils



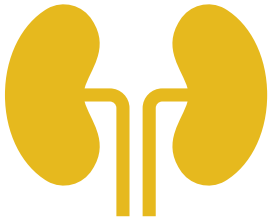
Nutrient Deficiencies

Macro Nutrients:

Energy – (factor in disease related factors)

Protein

Fluid (increased requirements)



Micronutrients:

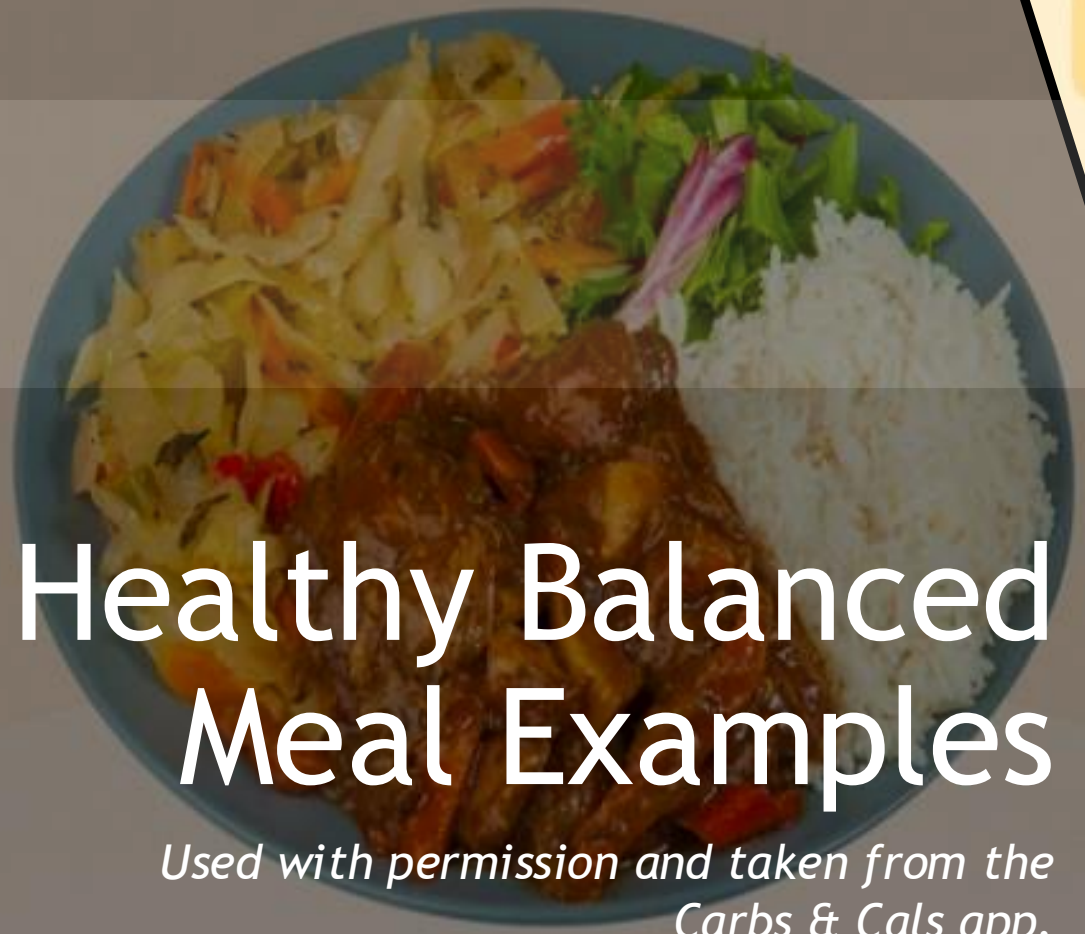
Zn, Se, Vit D, A, C, E, B vitamins, folic acid

Iron – (absence of iron overload)

Omega 3 Fatty acids

Healthy Balanced Meal Examples

Used with permission and taken from the Carbs & Cals app.



SICKLE CELL NUTRITION EATWELL GUIDE

Tailoring diet and nutrition knowledge and information to Sickle Cell Disease by increasing understanding of cultural and more sustainable foods enabling patients to better balance their diet and enhance health outcomes

Provide energy, carry fat and help absorb fat soluble vitamins

Fats and Oils

Choose unsaturated oils and use in small amounts

Best source of energy, fibre, B-vitamins

Starchy carbohydrates and wholegrains

SOURCE: 1. Public Health England in association with the Welsh Government, Food Standards Scotland, and the Foods Standards Agency, in Northern Ireland. 2. The Diverse Nutrition Association (African and Caribbean Eatwell Guide).

Choose wholegrain carbohydrates!
Good source of fibre to help
reduce and prevent constipation

SICKLE CELL NUTRITION EATWELL GUIDE

Tailoring diet and nutrition knowledge and information to Sickle Cell Disease by increasing understanding of cultural and more sustainable foods enabling patients to better balance their diet and enhance health outcomes

Provides vitamins and minerals, fibre and anti-oxidant rich foods

Vitamin A: dark green veg, yellow-orange veg, tomatoes, watermelon, apricots, peaches

Vitamin C: broccoli, green leafy veg, peppers, citrus fruits, berries, cantaloupe, strawberries

Vitamin E: Avocado and Vegetables, mangoes, spinach, red bell peppers, vegetables oils)

Fruit and Vegetables – 5 A Day

SOURCE: 1. Public Health England in association with the Welsh Government, Food Standards Scotland, and the Foods Standards Agency, in Northern Ireland. 2. The Diverse Nutrition Association (African and Caribbean Eatwell Guide).

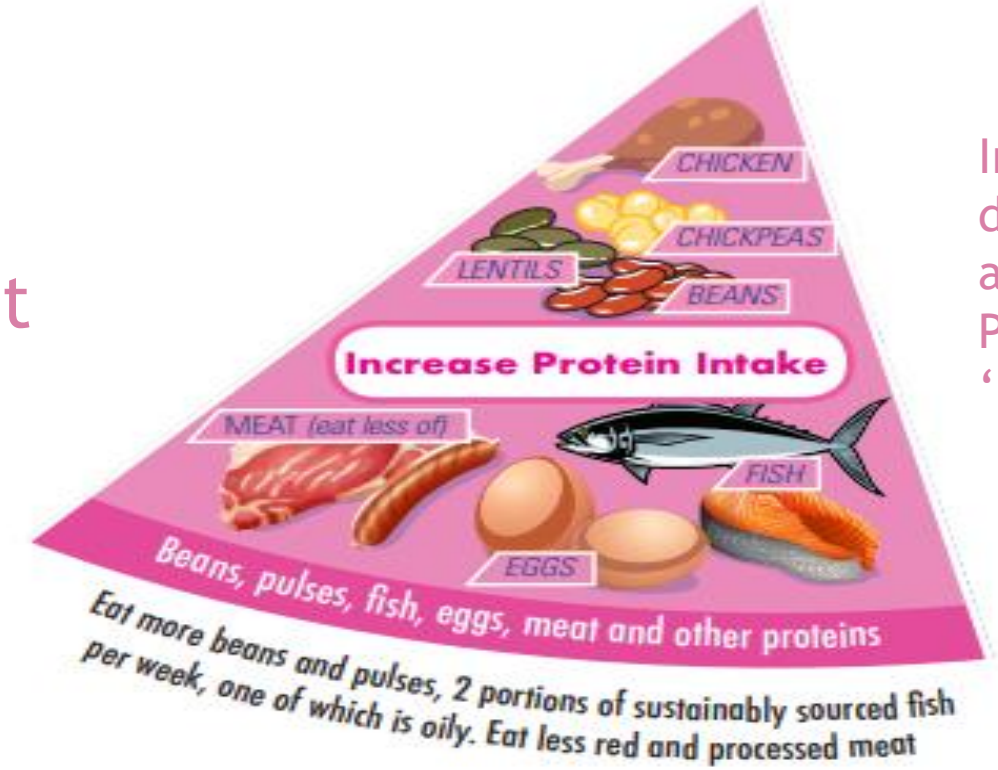

Choose a variety of fruit
and veg – Remember your 5
A day!

Folic Acid – Food Sources

How can you increase my daily folic acid intake?

- green leafy veg like spinach, kale, cabbage, spring greens, and broccoli
- cruciferous vegetables like brussels sprouts and asparagus
- beans and peas like chickpeas, kidney beans, black-eyes peas and lentils
- citrus fruit, oranges and other citrus foods
- tropical fruits like papaya and bananas,
- fortified foods like breakfast cereals, breads, flours, cornmeal, pasta, rice, other grain products
- other foods like liver, eggs, dairy products, meat, poultry, shellfish, nuts, yeast beef extract, avocado and wholewheat

Protein and Plant Alternatives



Important for growth and development, good health and vitality:
Part of all tissues
'Protein sparing'

SOURCE: 1. Public Health England in association with the Welsh Government, Food Standards Scotland, and the Foods Standards Agency, in Northern Ireland. 2. The Diverse Nutrition Association (African and Caribbean Eatwell Guide)

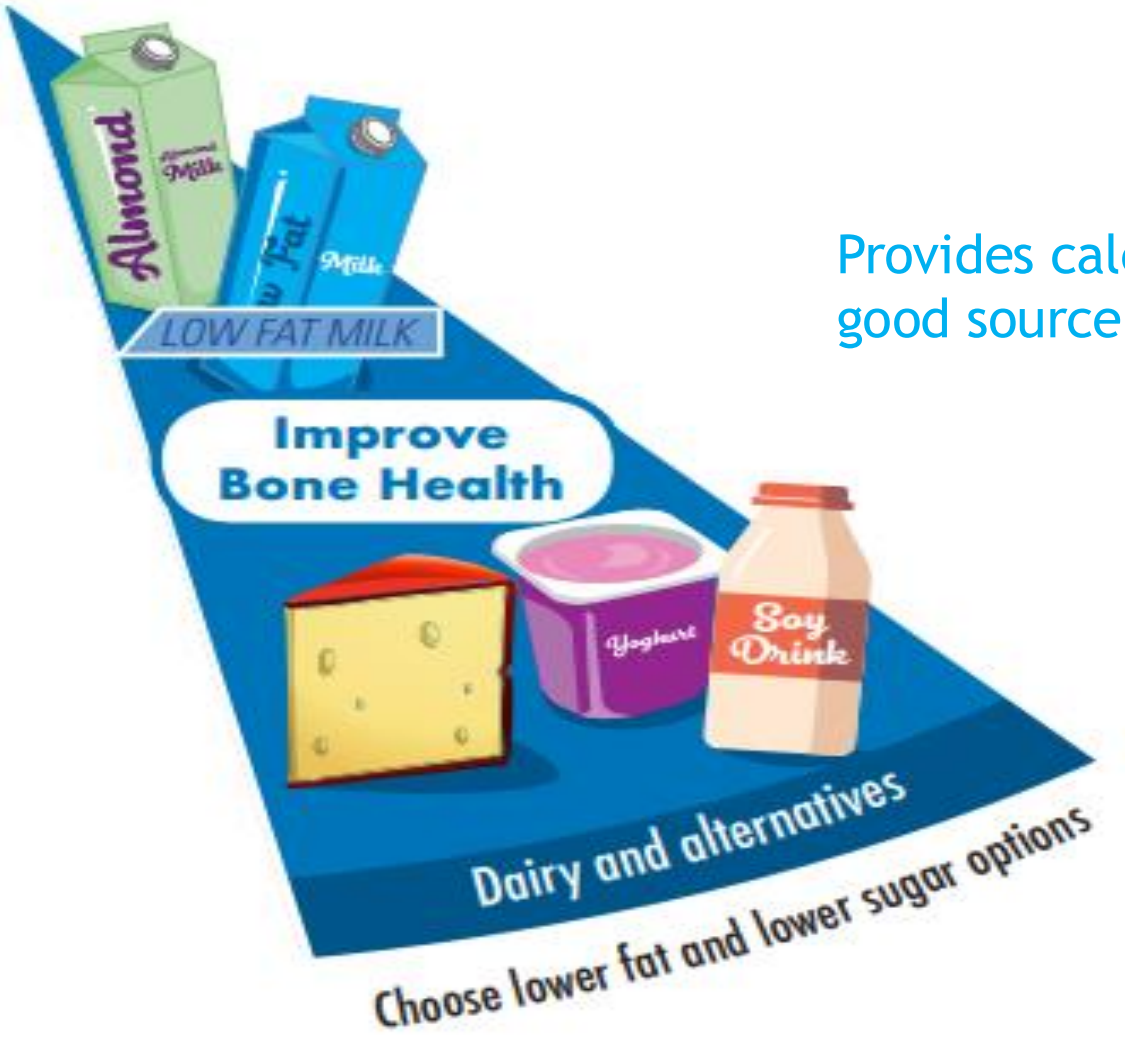
Protein sources -

Soya protein
Tofu
Tempeh
nuts and seeds

SICKLE CELL NUTRITION EATWELL GUIDE

Tailoring diet and nutrition knowledge and information to Sickle Cell Disease by increasing understanding of cultural and more sustainable foods enabling patients to better balance their diet and enhance health outcomes

Dairy and Alternatives



Provides calcium and a good source of protein

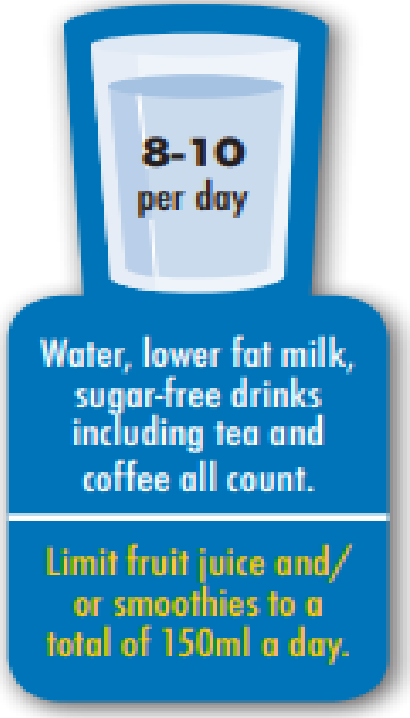
SOURCE: 1. Public Health England in association with the Welsh Government, Food Standards Scotland, and the Foods Standards Agency, in Northern Ireland. 2. The Diverse Nutrition Association (African and Caribbean Eatwell Guide).

Provide calcium and protein

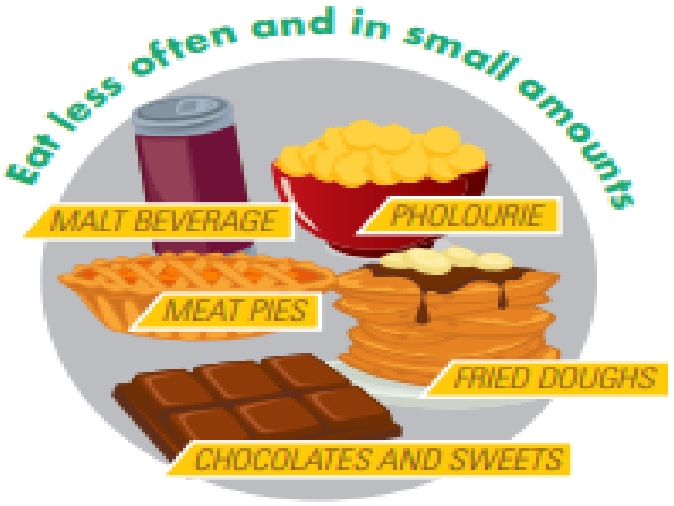
SICKLE CELL NUTRITION EATWELL GUIDE

Tailoring diet and nutrition knowledge and information to Sickle Cell Disease by increasing understanding of cultural and more sustainable foods enabling patients to better balance their diet and enhance health outcomes

PREVENT DEHYDRATION

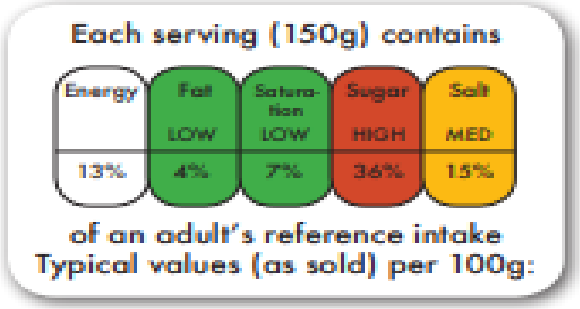


Additional Health Principles



CHOOSE HEALTHY OPTIONS

Use the traffic light system



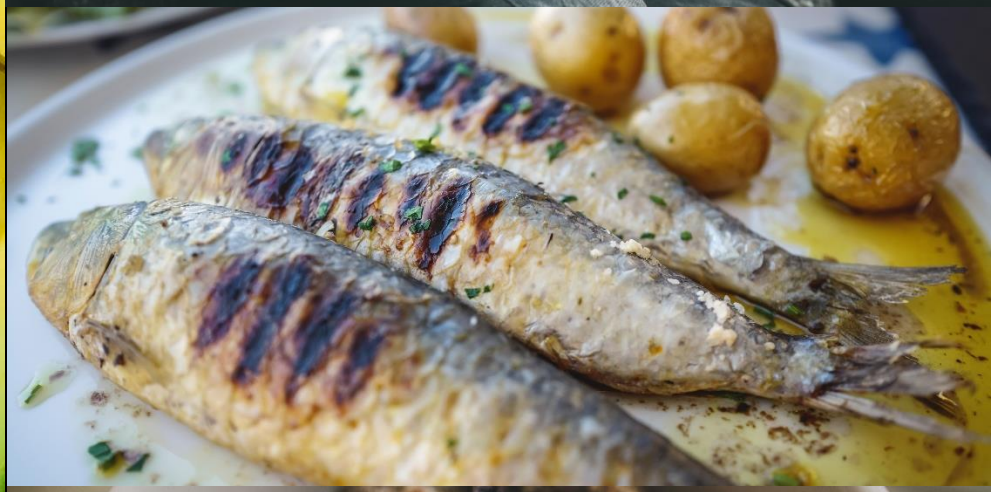
Choose foods lower in fat, salt and sugars

Essential for all the bodies processes

Make healthy dietary choices to maintain a healthy weight

SOURCE: 1. Public Health England in association with the Welsh Government, Food Standards Scotland, and the Foods Standards Agency, in Northern Ireland. 2. The Diverse Nutrition Association (African and Caribbean Eatwell Guide).

Healthy Fats – avocado, nuts and seeds, omega 3 fatty acids



Cutting down on sugar

Regularly consuming foods and drinks high in sugar increases your risk of obesity and tooth decay. Ideally, no more than 5% of the energy we consume should come from free sugars*. Currently, children and adults across the UK are consuming 2-3 times that amount.

Age	Recommended maximum free sugars intake	Sugar cubes
4-6 years	No more than 19g/day	5 cubes
7-10 years	No more than 24g/day	6 cubes
From 11 years, including adults	No more than 30g/day	7 cubes

Part 4: Benefit of good nutrition

Benefits Of Good Nutrition!

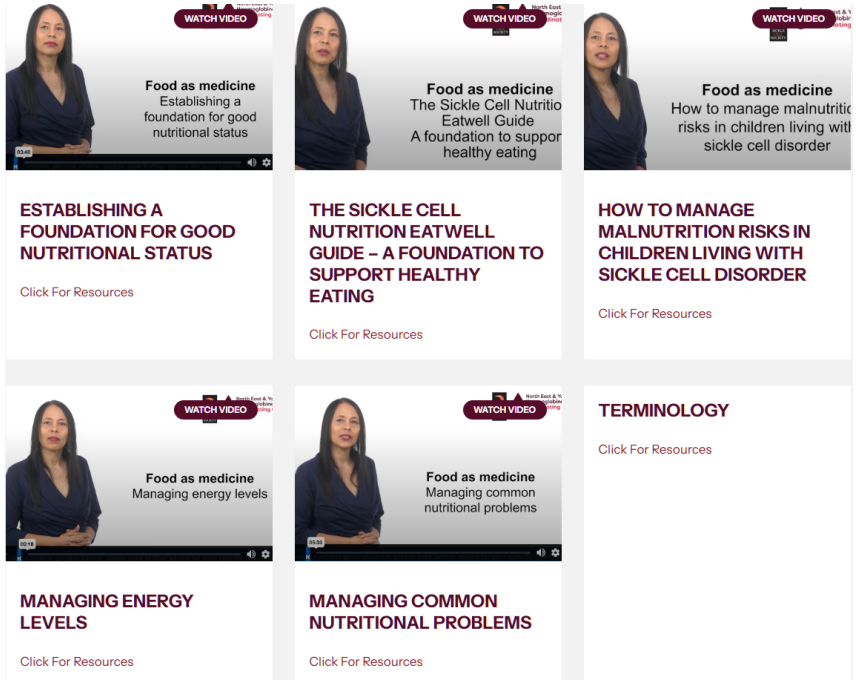
- **Improve your Nutritional Status by eating a Good balanced Diet to,**
  - Help you stay healthy for longer
  - Help your medication work better
  - Potentially improve the morbidity linked to SCD
  - Potentially reduce the impact of the side effects of medical treatment
  - Potentially reduce the frequency and intensity of a crisis
  - Help reduce length of stay in hospital
  - Help to increase recovery rates
  - Reduce the amount of medication to manage SCD

One thing you will

- Take Away…

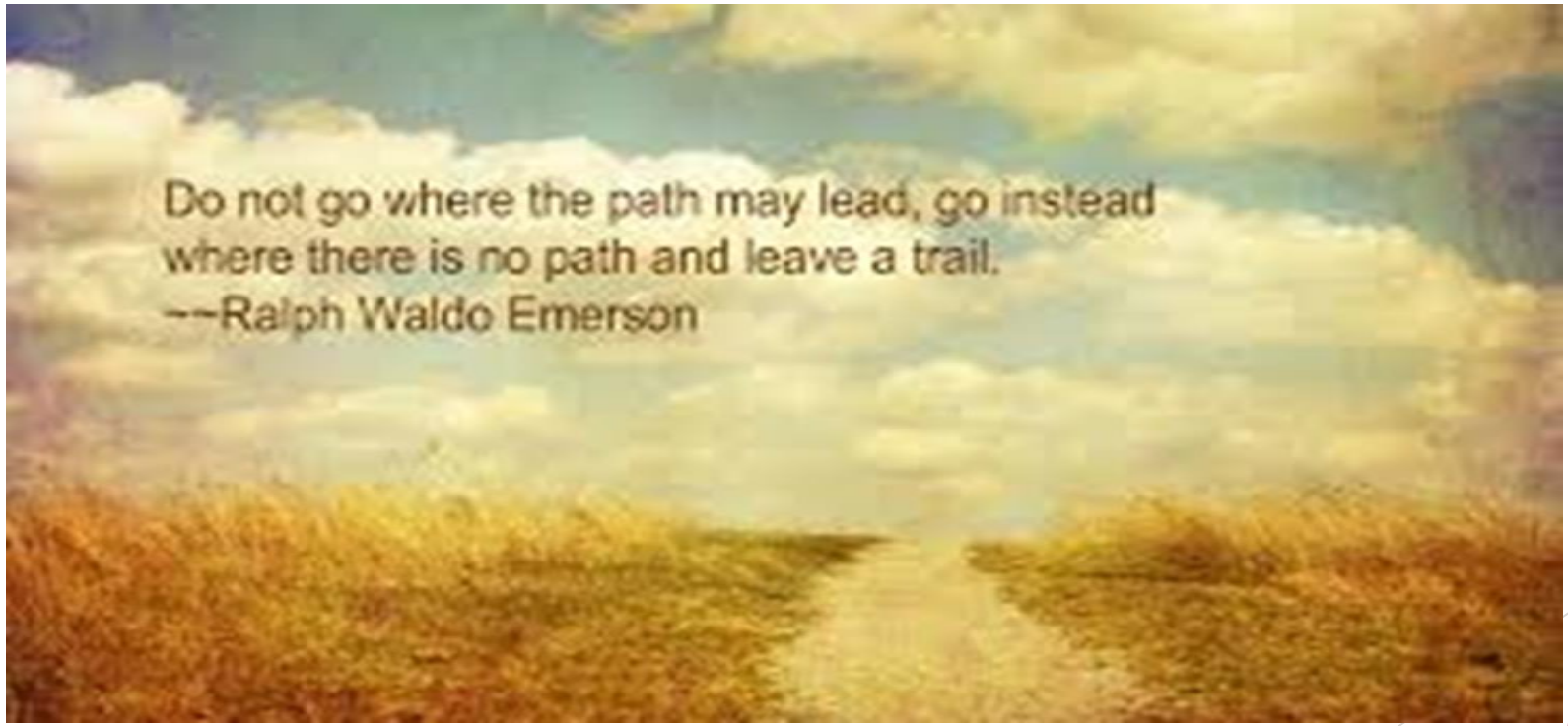
Videos

Collaboration with NE & Y HCC and Sickle Cell Society:
https://www.ney-hcc.co.uk/resources/



Thank You!

"Every closed door brings you closer to your Destiny"
Dr Claudine Matthews



North East & Yorkshire Haemoglobinopathy Coordinating Centre

New Treatments in Sickle Cell Disorder

Dr Clare Samuelson

Outline of Talk

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Why do we need new treatments?
- Bone marrow transplant
- Gene therapy
- Clinical trials
- Discussion and questions

Why Do We Need New Treatments?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Hydroxycarbamide is a good treatment for most people
- Some need regular transfusions
- But…. many they still experience pain and other problems
- Some people can’t receive these treatments
- Limited options for alternative treatments

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Bone Marrow Transplant

What is it and who is it for?

Bone Marrow Transplant

North East & Yorkshire Haemoglobinopathy Coordinating Centre

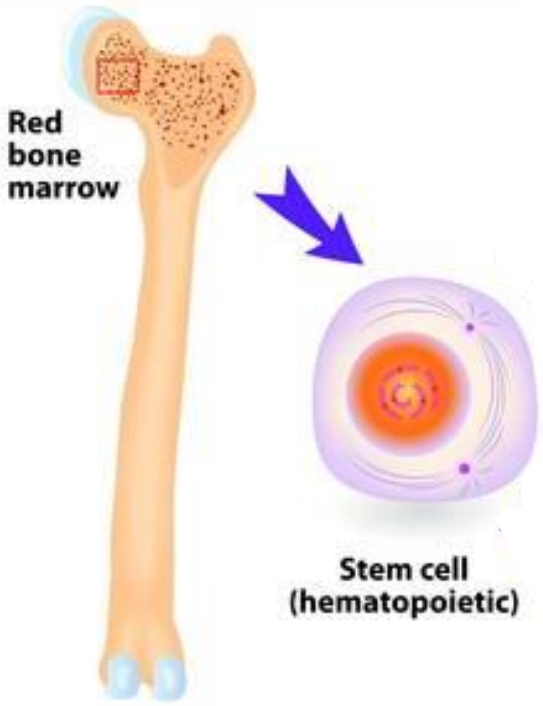
Also called:

- Haematopoietic stem cell transplant
- Stem cell transplant
- BMT
- HSCT

New for adults (since 2019)

How Does a Bone Marrow Transplant Work?

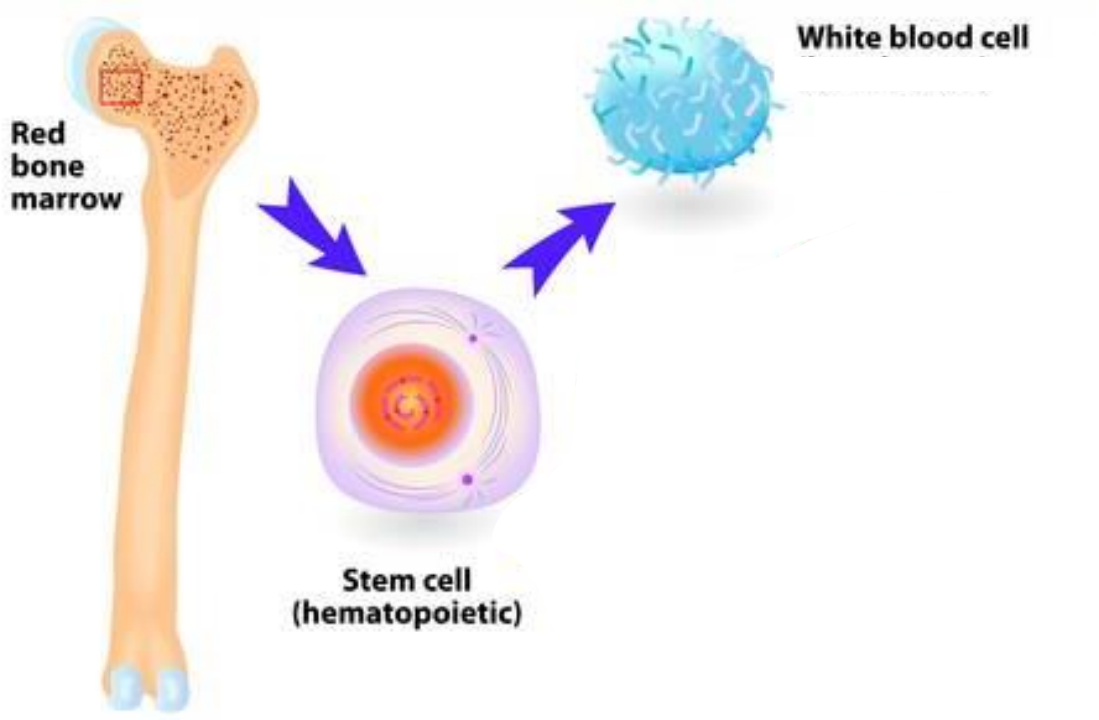
North East & Yorkshire Haemoglobinopathy Coordinating Centre



Stem cells in your bone marrow make all your other blood cells

How Does a Bone Marrow Transplant Work?

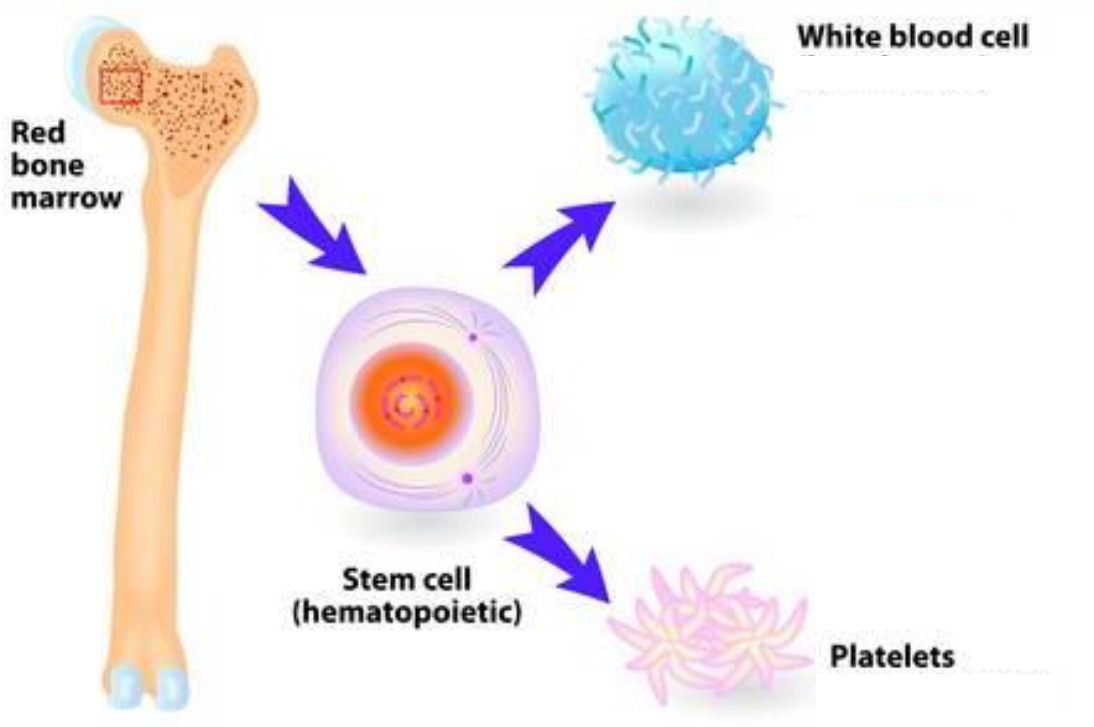
North East & Yorkshire Haemoglobinopathy Coordinating Centre



Stem cells in your bone marrow make all your other blood cells

How Does a Bone Marrow Transplant Work?

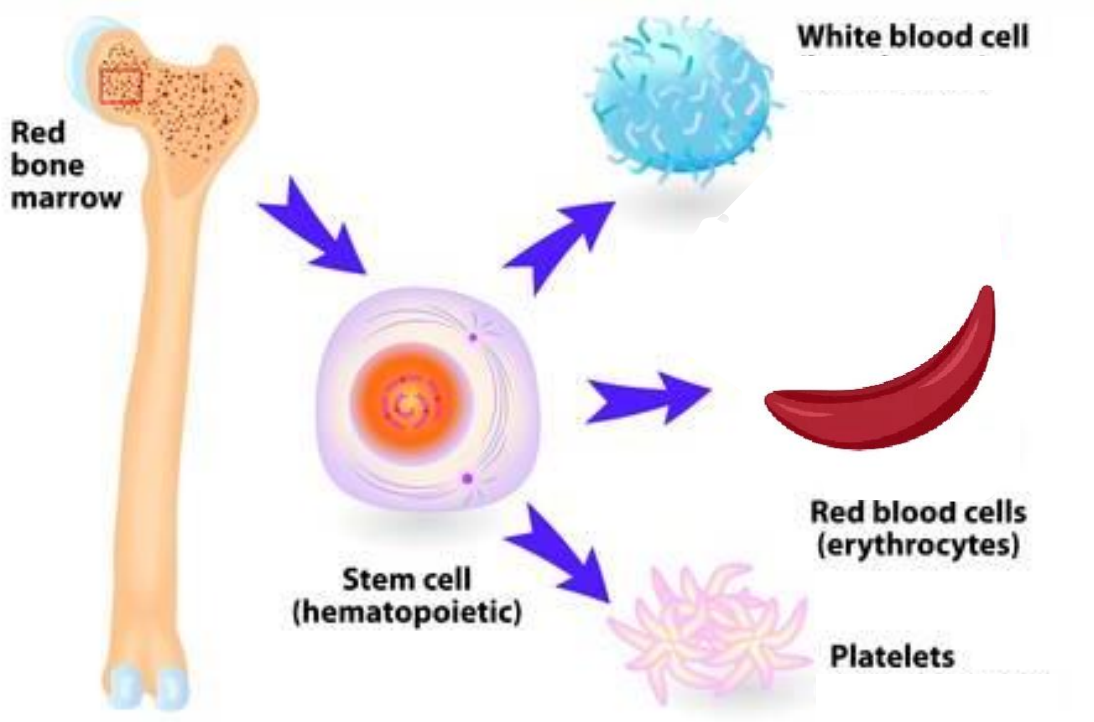
North East & Yorkshire Haemoglobinopathy Coordinating Centre



Stem cells in your bone marrow make all your other blood cells

How Does a Bone Marrow Transplant Work?

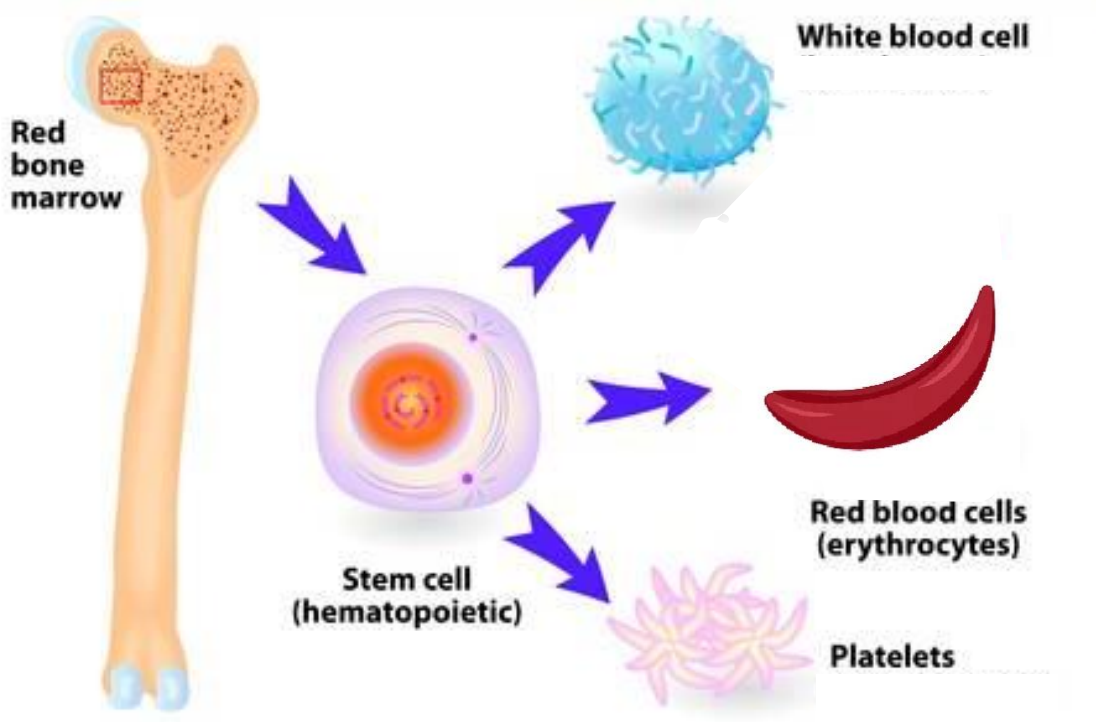
North East & Yorkshire Haemoglobinopathy Coordinating Centre



Stem cells in your bone marrow make all your other blood cells

How Does a Bone Marrow Transplant Work?

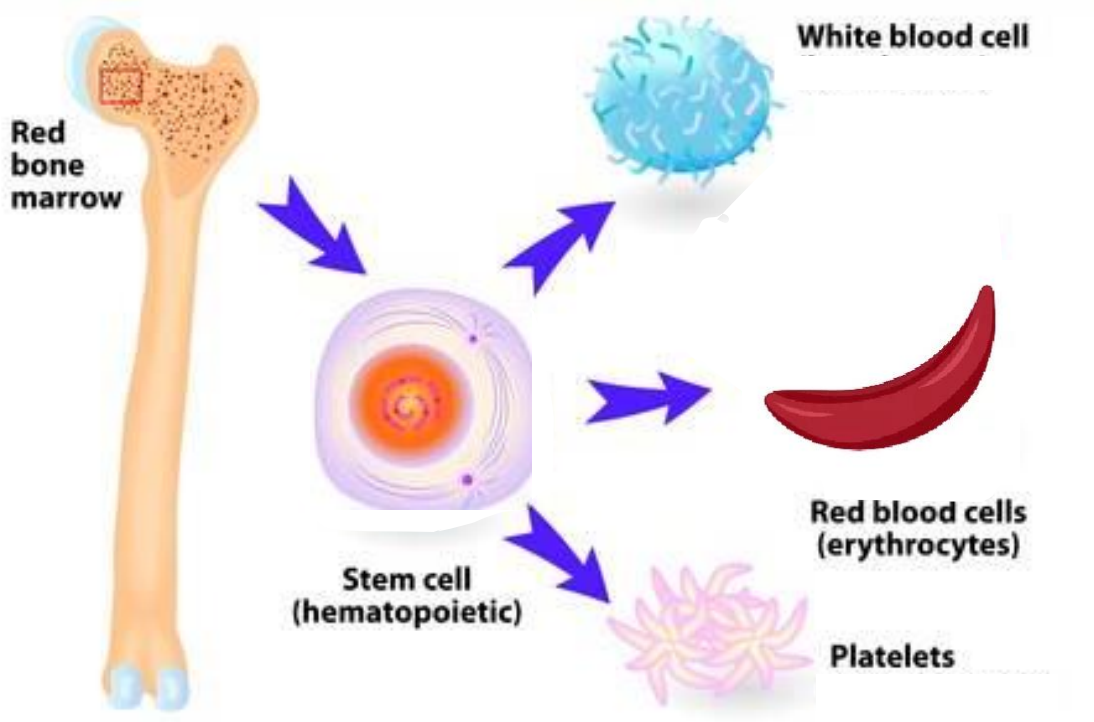
North East & Yorkshire Haemoglobinopathy Coordinating Centre



Sickle cell is caused by a genetic change in your stem cells

How Does a Bone Marrow Transplant Work?

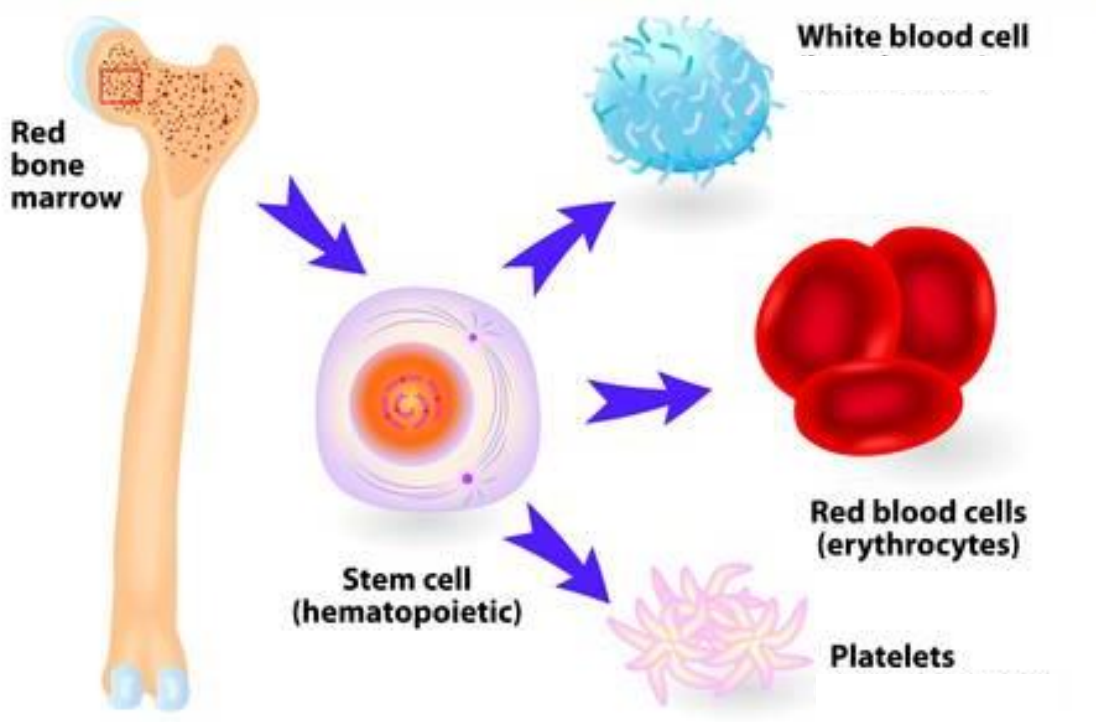
North East & Yorkshire Haemoglobinopathy Coordinating Centre



If we can replace your stem cells with healthy stem cells from a donor…

How Does a Bone Marrow Transplant Work?

North East & Yorkshire Haemoglobinopathy Coordinating Centre



This can cure sickle cell if the transplant is successful

What are the Downsides to Bone Marrow Transplant?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Chemotherapy (or similar) to remove your own stem cells

- Time in hospital, sometimes long recovery
- Risk of damage to organs
- Affects fertility (options available to help)

Safest in young children – less safe for older people

Risk that donor cells attack your body *(graft versus host disease)*

Does not always work

Who Might be Offered Bone Marrow Transplant? **Children**

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Severe sickle cell disorder:

- Still having frequent crises even with hydroxycarbamide (4 or more per year needing to come to hospital or affecting schooling)
- 2 or more chest crises even with hydroxycarbamide
- Sickle cell has caused a stroke or other problems with brain health

Donor: matched healthy family member or can sometimes use half-match family member or unrelated donor

Who Might be Offered Bone Marrow Transplant? **Adults**

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Severe sickle cell disorder:

- 3 or more severe crises per year despite hydroxycarbamide
- 2 or more chest crises despite hydroxycarbamide
- Needing regular blood transfusions
- Sickle cell has caused organ damage such as stroke

Fit enough to receive the transplant

Fully matched family donor (brother or sister)

REDRESS Clinical Trial

North East & Yorkshire Haemoglobinopathy Coordinating Centre

REDRESS

- Clinical trial
- Open in Sheffield for people in NE&Y
- For adults who need a transplant but don't have a matched family donor
- 'Haploidentical' = half matched family members as donors
- 50 / 50 chance : transplant or standard treatment

What is it Like for the Donor?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

1. Blood test (usually) to see if they are a match
2. Checks to make sure they are well and can donate safely
3. Injections to stimulate the stem cells for a few days
4. Stem cells collected from the vein, or from the hip bones (a short procedure under anaesthetic)

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Gene Therapy

What is it and who is it for?

Gene Therapy

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Also called:

- Genome editing
- Exa-cel
- Casgevy®

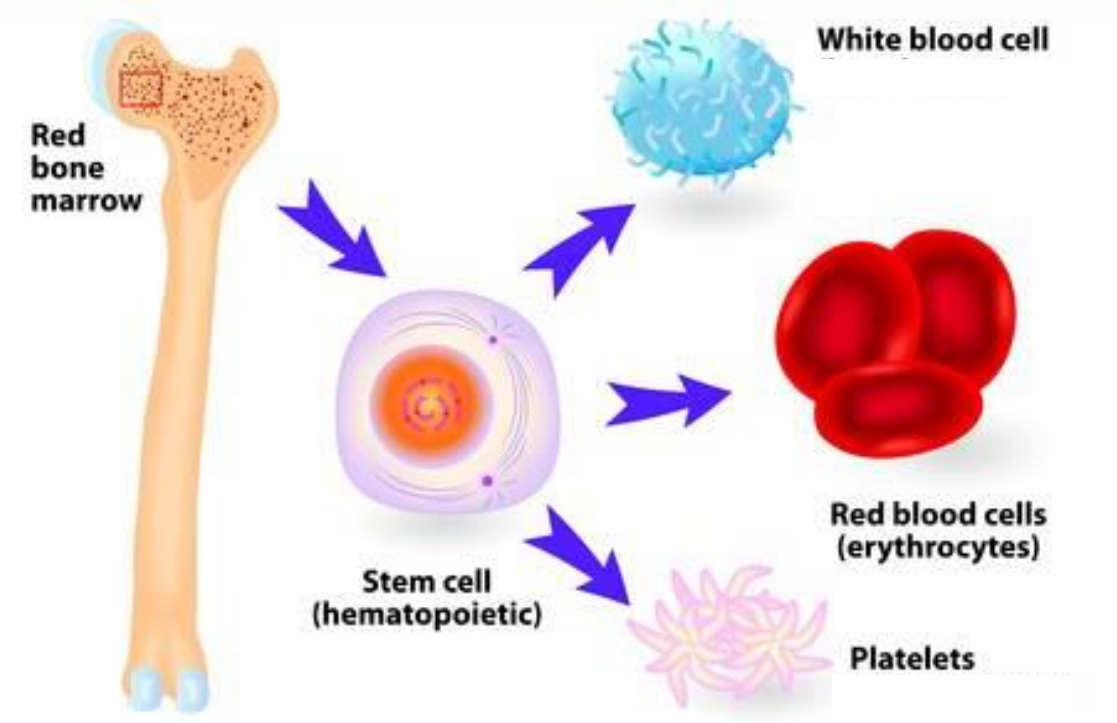
Available since January 2025 for people aged 12 years +

How Does Gene Therapy Work?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Uses your own stem cells

Alters your genes to make more fetal haemoglobin (HbF)



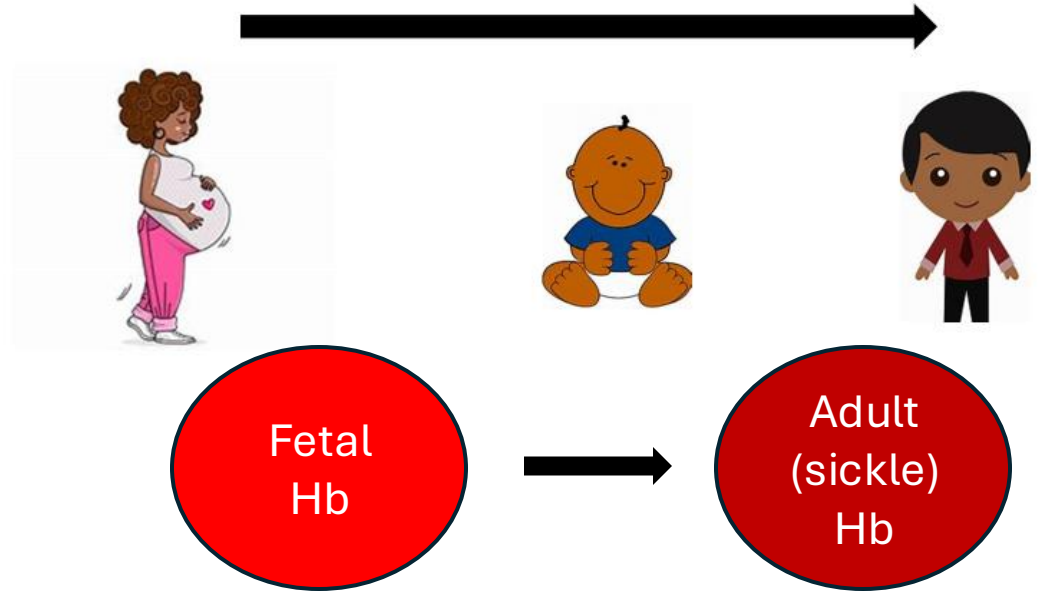
How Does Gene Therapy Work?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Uses your own stem cells

Alters your genes to make more fetal haemoglobin (HbF)

We usually stop making fetal Hb after birth



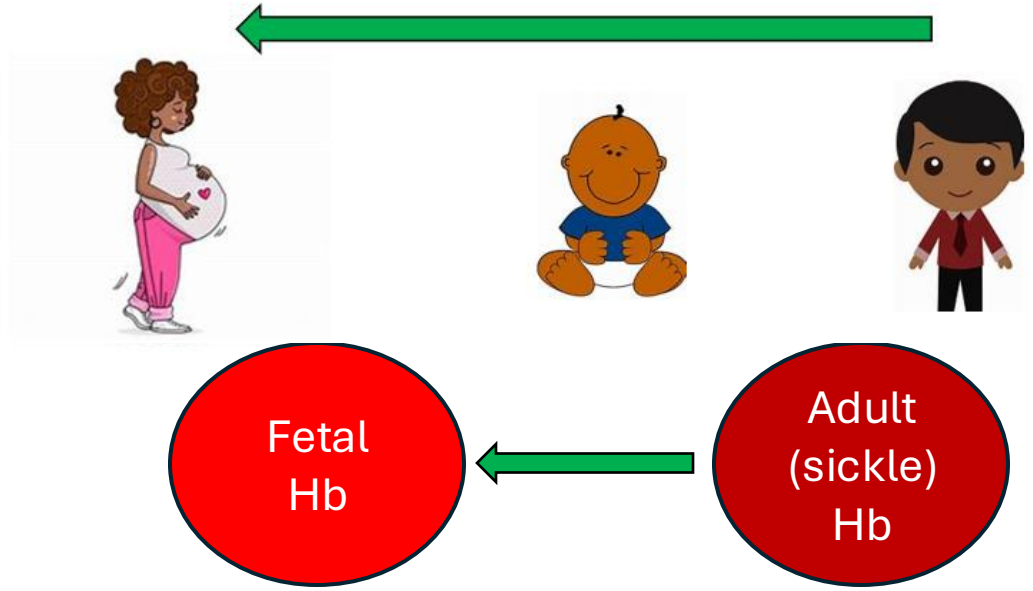
How Does Gene Therapy Work?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Uses your own stem cells

Alters your genes to make more fetal haemoglobin (HbF)

Genome editing switches fetal Hb back on



How Does Gene Therapy Work?

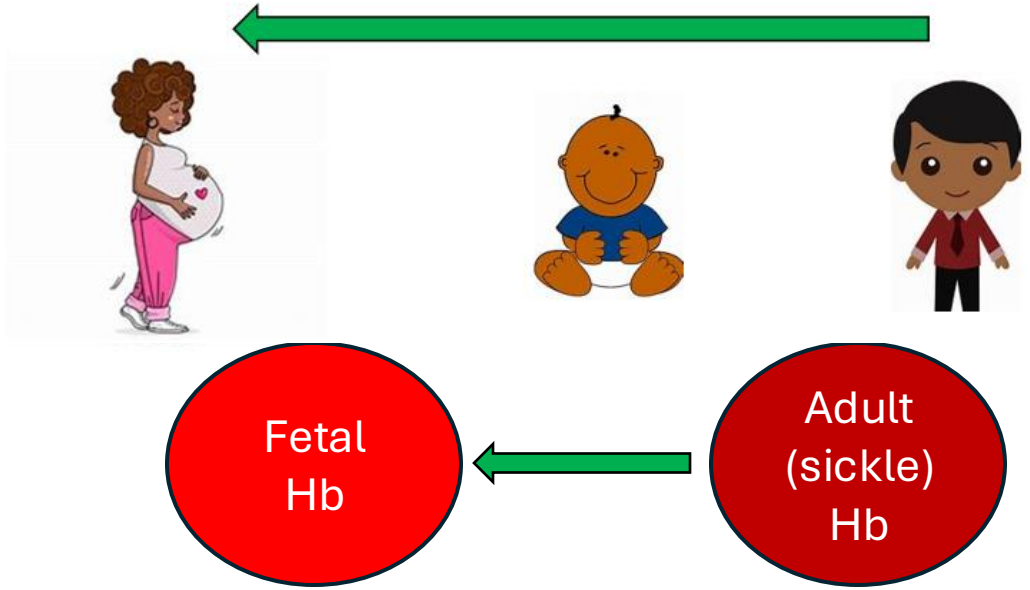
North East & Yorkshire Haemoglobinopathy Coordinating Centre

Uses your own stem cells

Alters your genes to make more fetal haemoglobin (HbF)

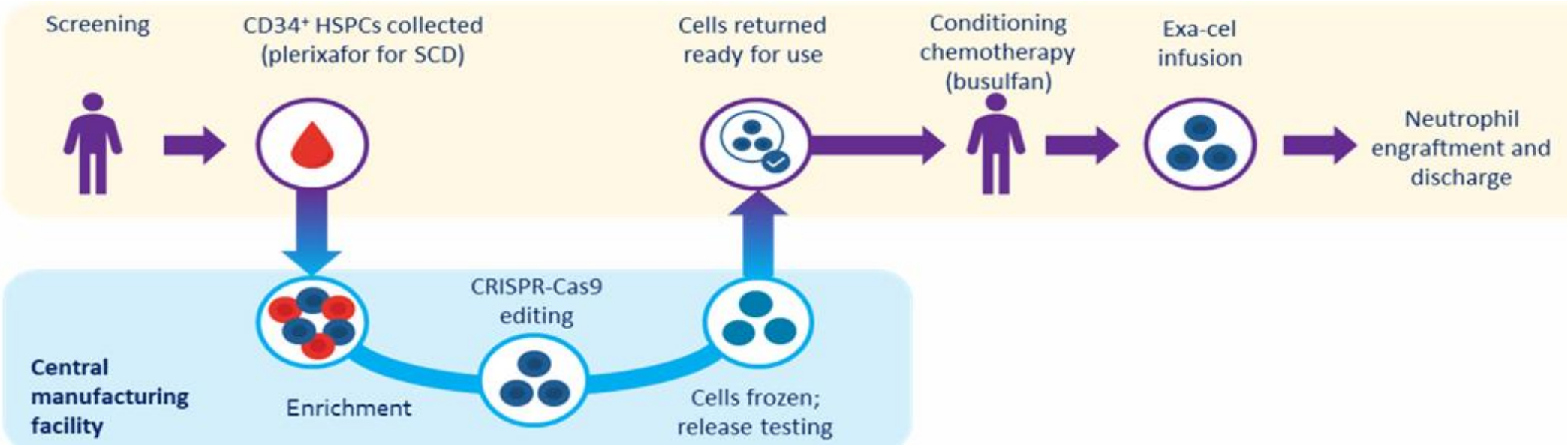
Genome editing switches fetal Hb back on

More HbF helps to prevent crises



What is it Like to have Gene Therapy?

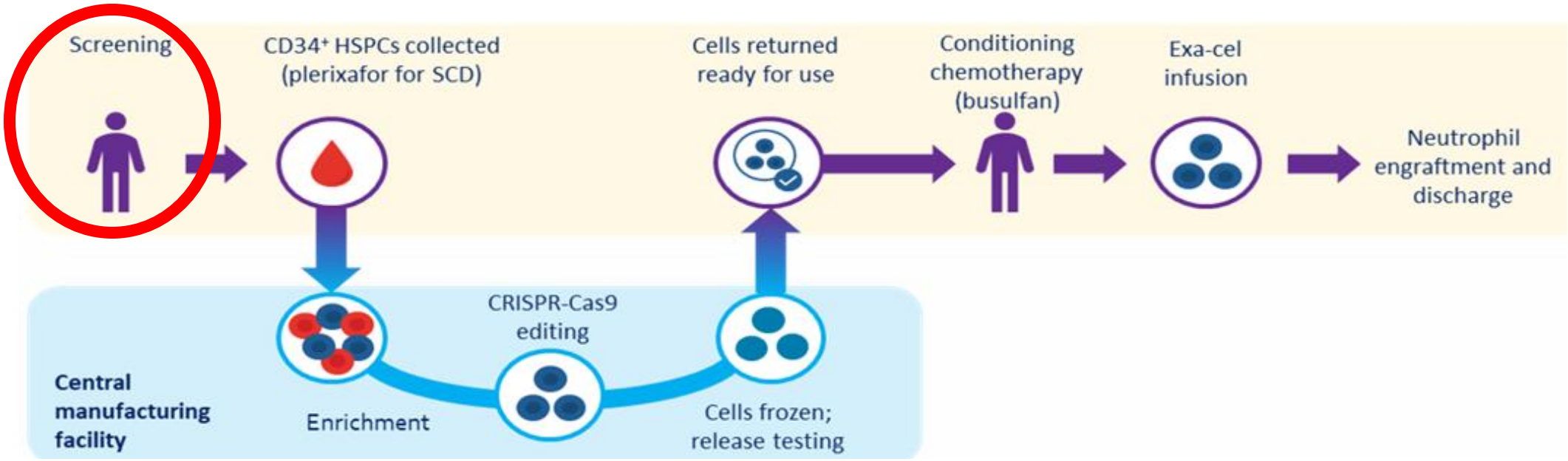
North East & Yorkshire Haemoglobinopathy Coordinating Centre



Frangoul et al, 2024

What is it Like to have Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre



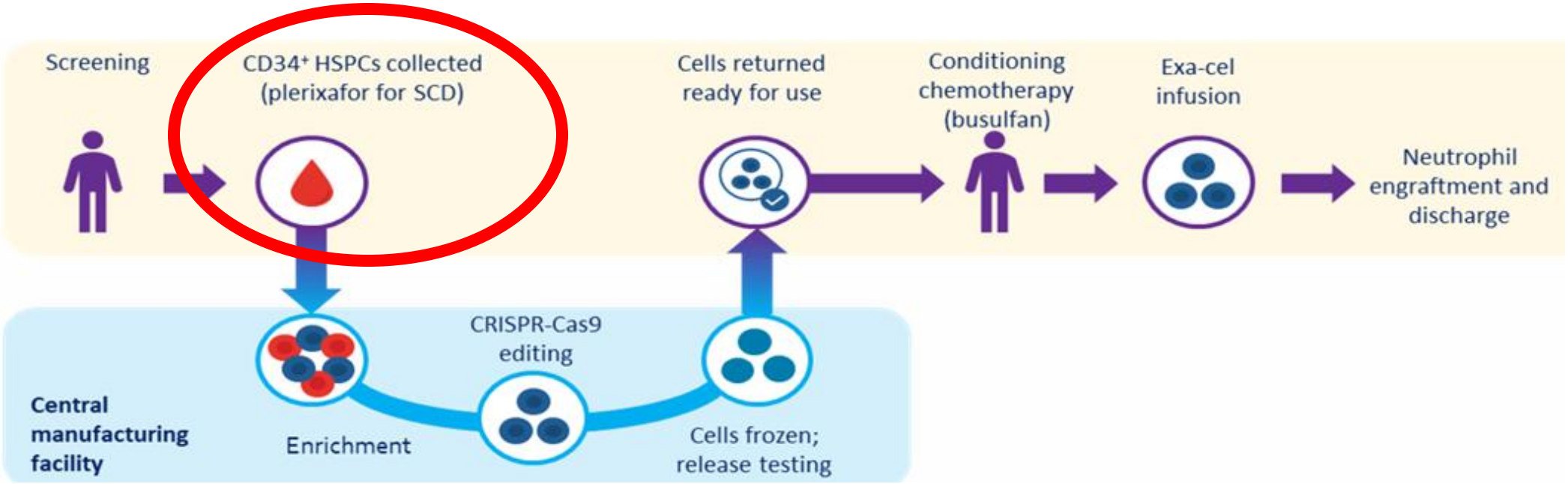
Screening: making sure the treatment is suitable for you

- Check you don't have a matched family donor for bone marrow transplant
- Review how often you have crises and any other complications
- Check you are fit enough to manage the treatment
- Making sure you have all the right information to make a decision

Frangoul et al, 2024

What is it Like to have Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre



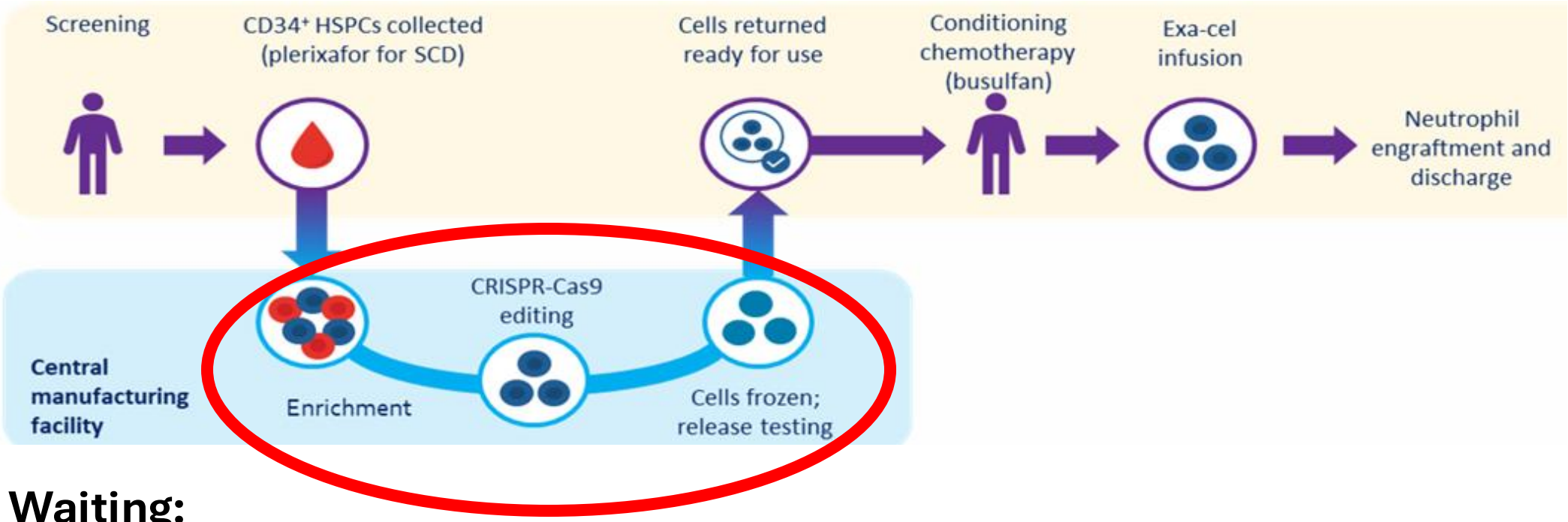
Collecting your stem cells:

- Injection to stimulate your stem cells to come out of bone marrow
- Stem cells collected from your vein (like red cell exchange)
- Often takes multiple times to successfully collect enough

Frangoul et al, 2024

What is it Like to have Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre



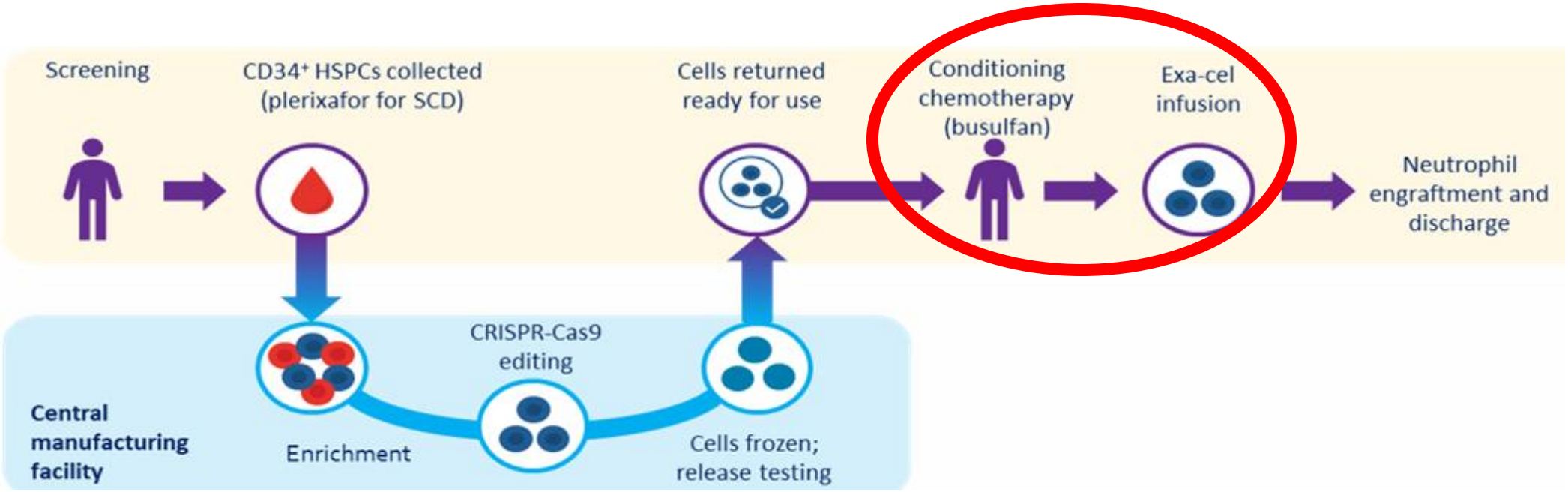
Waiting:

- Wait usually 6 months or could be longer
- Keep going with your normal treatment
- Fertility preservation might take place

Frangoul et al, 2024

What is it Like to have Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre



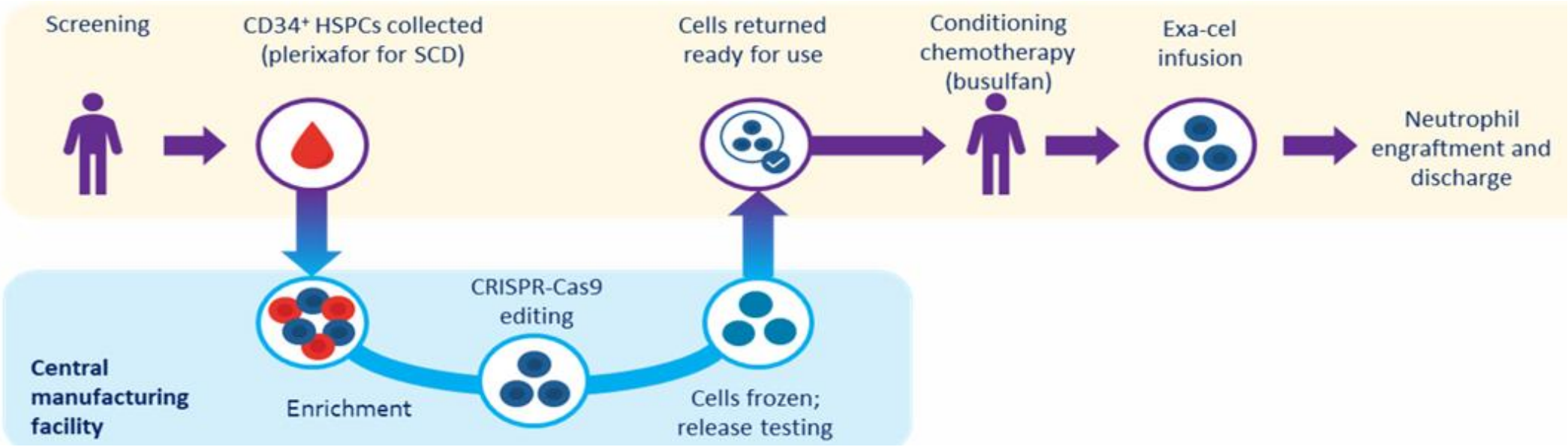
Receiving the modified stem cells:

- Stay in hospital usually 4-6 weeks
- Strong chemotherapy given then the Exa-cel (stem cells)
- Wait until your blood counts recover then you can go home

Frangoul et al, 2024

What is it Like to have Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre



Recovery:

- See the specialist team regularly
- Probably 6 – 12 months for complete recover (think about work or study requirements – can you do some from home? Financial support?)

Frangoul et al, 2024

What are the Benefits of Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Most people who had gene therapy on the clinical trial did not have crises after treatment
  - 43/45 people had no pain for at least 12 months
  - 45/45 people had at 12 months + out of hospital
  - After treatment crises might still happen especially with triggers such as infection or surgery
- One-off treatment with probably long-term effect

CLIMB SCD-121 and CLIMB-131 clinical trials

Vertex, EHA 2025

What are the Downsides of Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Strong chemotherapy
  - Time in hospital, sometimes long recovery
  - Risk of damage to organs / life-threatening complications
  - Affects fertility (options available to help)
- Only currently available for age 12 years + (Clinical trial looking at treatment for younger children)
- First person treated 6 years ago – we don't have long term data

Who Might be Offered Gene Therapy?

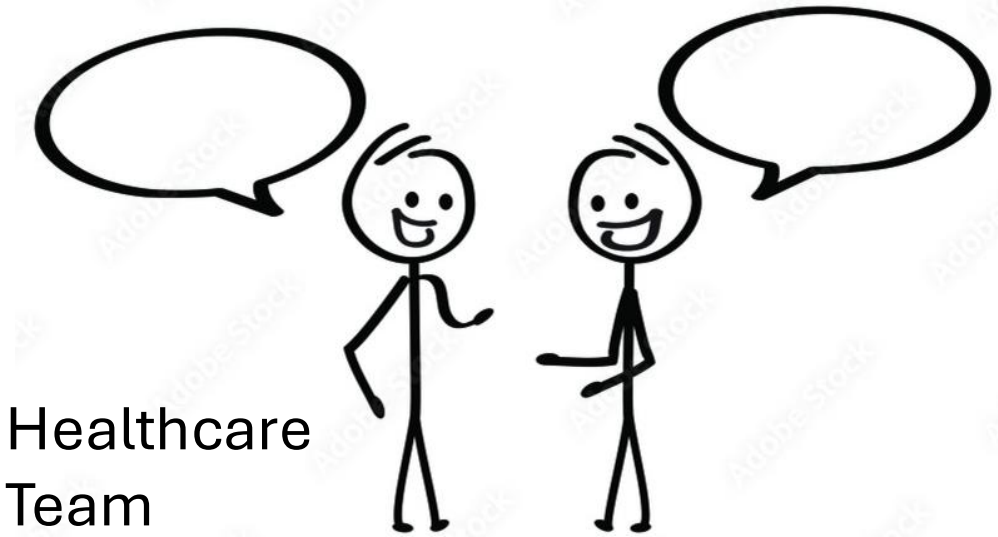
North East & Yorkshire Haemoglobinopathy Coordinating Centre

- Severe sickle cell disorder:
  - Frequent crises needing medical attention (2 or more per year for 2 years)
- No matched family donor
- Age 12+ (for older adults needs careful consideration)

Gene therapy treatment currently delivered in Manchester for North of England

Where Can I Find More Information about Bone Marrow Transplant and Gene Therapy?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

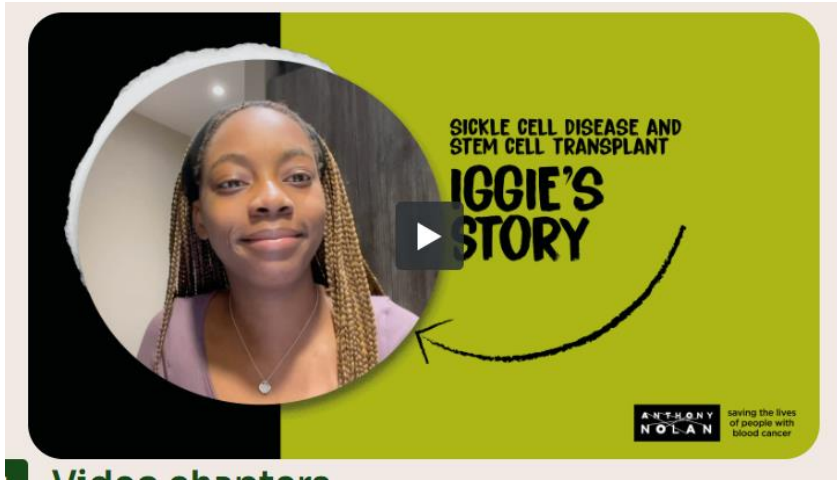


Healthcare Team

SICKLE CELL SOCIETY

ANTHONY NOLAN

Saving lives through stem cells



North East & Yorkshire Haemoglobinopathy Coordinating Centre

Clinical Trials

Why do We Need Clinical Trials in Sickle Cell?

North East & Yorkshire Haemoglobinopathy Coordinating Centre

- We need more treatments for sickle cell:
  - Only 4 new treatments approved for sickle cell in the UK in over 20 years – 2 since withdrawn
  - Transplant and gene therapy are not right for everyone
  - More choices and better treatment options needed
- Clinical trials can offer access to new (experimental) treatments
- Clinical trials are the only way to gain more knowledge and better treatment options

Clinical Trials in the NE&Y Region
https://www.ney-hcc.co.uk/research-projects/

North East & Yorkshire Haemoglobinopathy Coordinating Centre

FLORAL

SHEFFIELD TEACHING HOSPITALS NHS FOUNDATION TRUST

EXPLORE TRIAL

INSCHOOL

LEEDS TEACHING HOSPITALS NHS TRUST

EXPLORE TRIAL

HAEMATOLOGY LIVED EXPERIENCE AND OUTCOMES (HALO)

SHEFFIELD TEACHING HOSPITALS NHS FOUNDATION TRUST, THE LEEDS TEACHING HOSPITALS NHS TRUST, THE NEWCASTLE UPON TYNE HOSPITALS NHS FOUNDATION TRUST

EXPLORE TRIAL

REDRESS (RELATED HAPLO-DONOR HAEMATOPOIETIC STEM CELL TRANSPLANTATION FOR ADULTS WITH SEVERE SICKLE CELL DISEASE)

SHEFFIELD TEACHING HOSPITALS NHS FOUNDATION TRUST

EXPLORE TRIAL

IMPROVING BLACK HEALTH OUTCOMES (IBHO) BIORESOURCE

LEEDS TEACHING HOSPITALS NHS TRUST, THE NEWCASTLE UPON TYNE HOSPITALS NHS FOUNDATION TRUST

EXPLORE TRIAL

NIHR RAPID SERVICE EVALUATION TEAM (REVAL)

LEEDS TEACHING HOSPITALS NHS TRUST, SHEFFIELD TEACHING HOSPITALS NHS FOUNDATION TRUST

EXPLORE TRIAL

HIBISCUS

SHEFFIELD TEACHING HOSPITALS NHS FOUNDATION TRUST

EXPLORE TRIAL

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Questions and Discussion

Close

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Feedback

QR codes on tables

Food Parcels

16:20	Newcastle Arrange to leave to be on coaches - collect food for journey home
16:30	Leeds Arrange to leave to be on coaches - collect food for journey home
16:40	Sheffield Collect food parcel