North East & Yorkshire Haemoglobinopathy Coordinating Centre

Sickle Cell Disorder

Patient Education Event

21st June 2025

North East & Yorkshire Haemoglobinopathy Coordinating Centre

What Is Sickle Cell Disease?

Dr Emma Astwood
Paediatric Haematologist
Sheffield Children's Hospital

North East & Yorkshire Haemoglobinopathy Coordinating Centre

To understand:

- What sickle cell disease is
- How sickle cell disease presents and why
- Possible complications of sickle cell disease
- How to manage sickle cell disease at home

What Is Blood?

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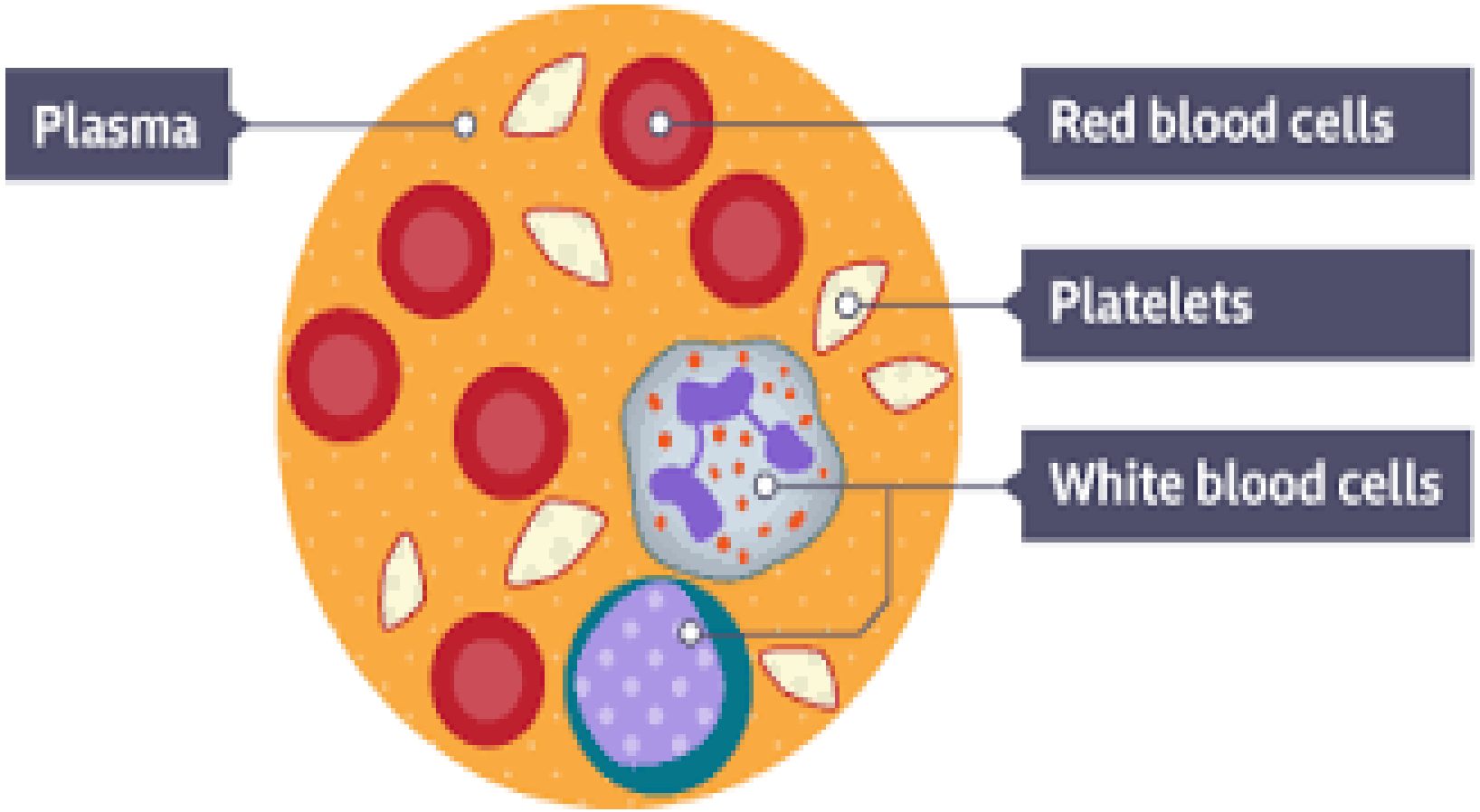

Red Blood Cells

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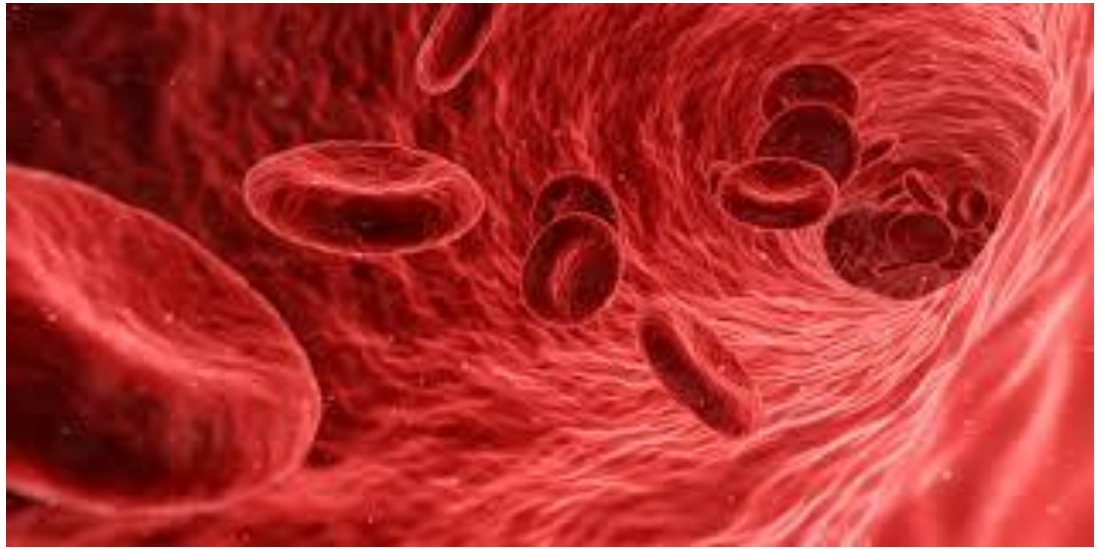
- Red Blood Cells are produced in the bone marrow in the centre of bones.
- Red blood cells carry oxygen from our lungs to the rest of the body and then carry carbon dioxide from the tissues back to the lungs as a waste product to be exhaled.
- Normal red blood cells live for 120 days and then get destroyed by the spleen.
- Normal red blood cells are soft and tiny and squeeze through tiny blood vessels and they look like a soft doughnut.



Haemoglobin

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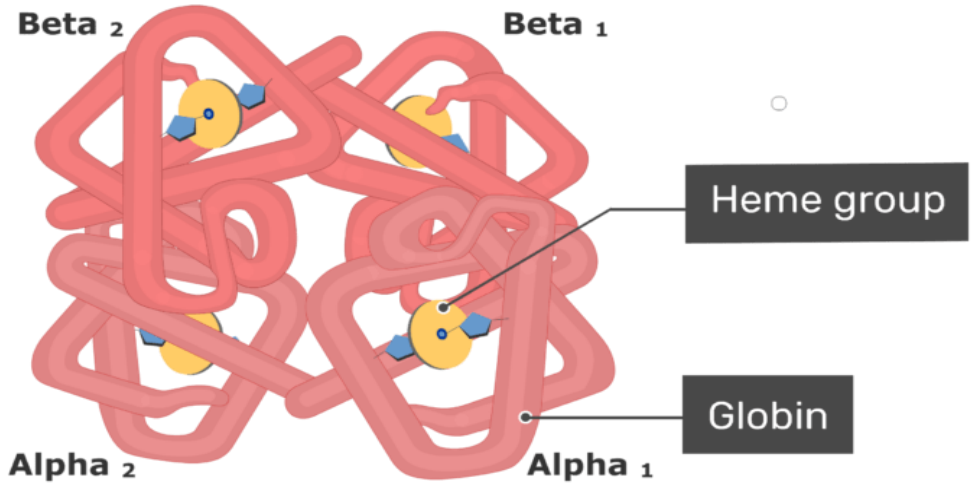
- Each red blood cell contains 260-280 million molecules of haemoglobin.
- Haemoglobin is a protein in the red blood cells which gives it its red colour and carries oxygen around the body.
- It has 2 parts – heme and globin
  - Heme – made up of iron (carries oxygen)
  - Globin – proteins which join to the haem molecules (surround and protect the haem molecules)
- There are different globin chains
  - Alpha, beta, gamma, delta, epsilon, zeta)
- The structure of each globin chain is genetically determined



What Is Sickle Cell Disease?

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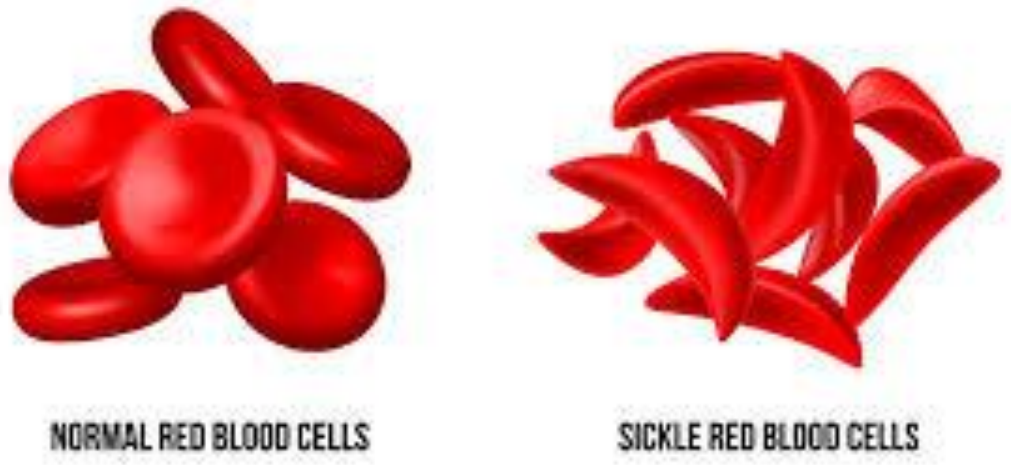
- A term covering a number of different but similar conditions that affect haemoglobin.
- Due to a genetic change on the β globin chain which leads to a lifelong disorder affecting the red blood cells, leading to abnormal haemoglobin and rigid, sickle-shaped cells.
- Types most commonly seen in UK:
  - Sickle Cell Anaemia (HbSS)
  - Sickle Haemoglobin C disease (HbSC)
  - Sickle Beta Thalassaemia (HbS/β thal)



Prevalence of Sickle Cell Disease

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- Most people affected are of African or African-Caribbean origin, although the sickle gene is found in all ethnic groups.
- It is estimated that there are 17,500 people with sickle cell disease in the UK and it is the most common genetic condition in the UK.
- The prevalence of the disease is increasing in the UK because of immigration and new births.

How Is Sickle Cell Disease Inherited?

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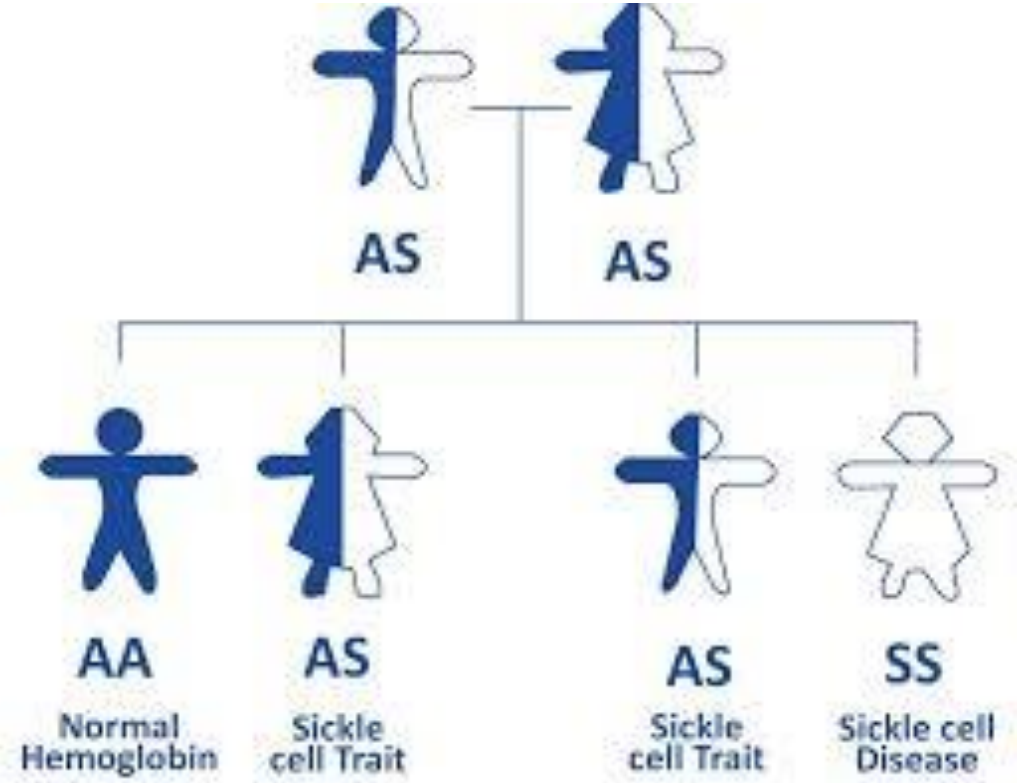

Why Does HbS Cause Problems?

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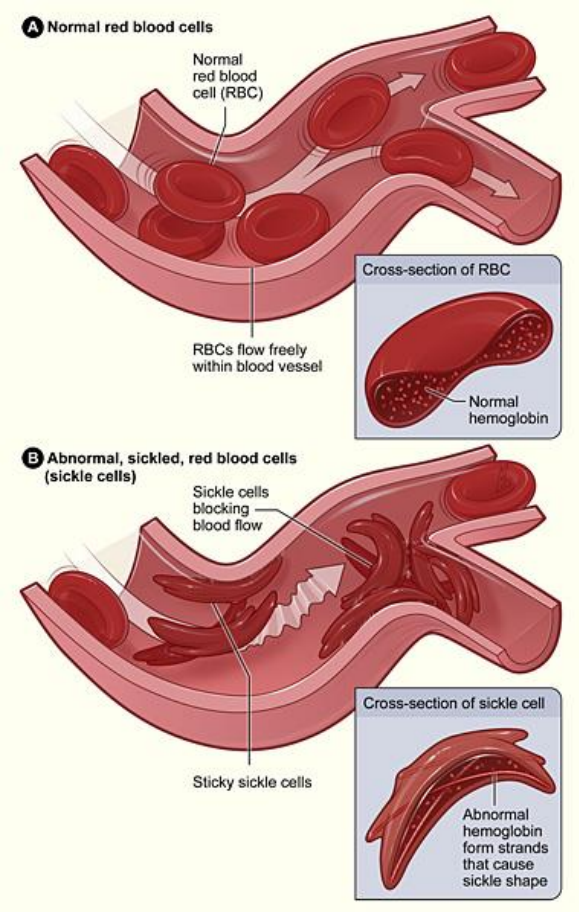

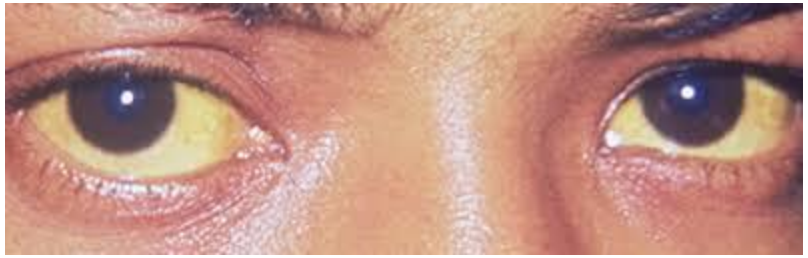

How Can Sickle Cell Disease Present?

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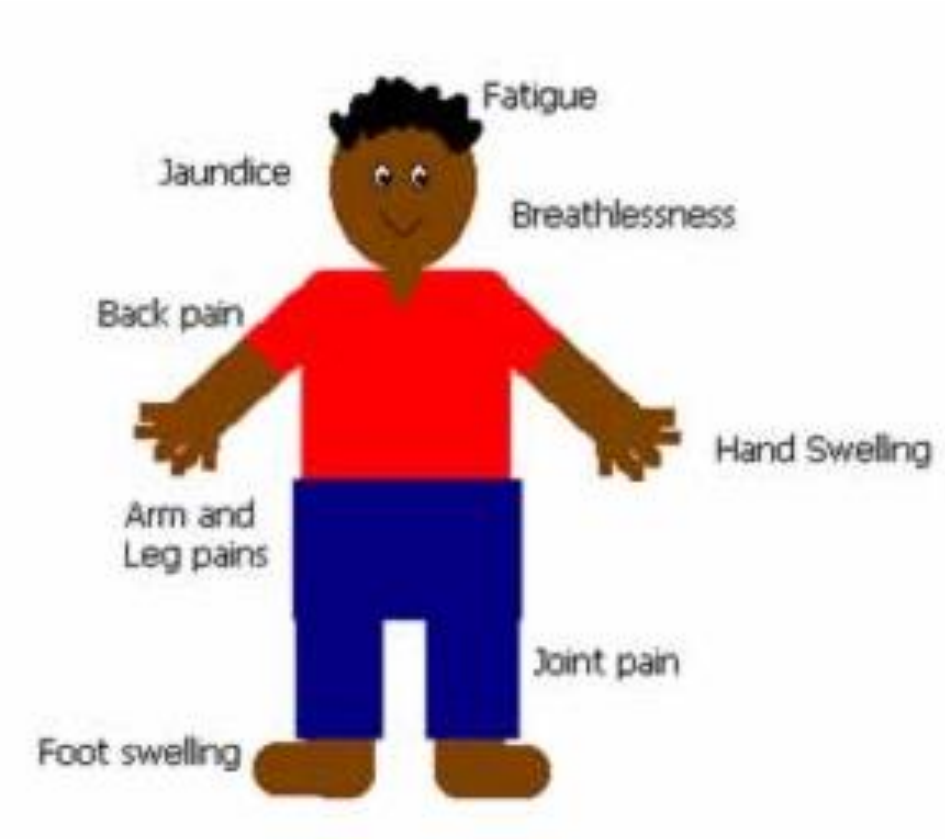

Potential Complications of Sickle Cell Disease

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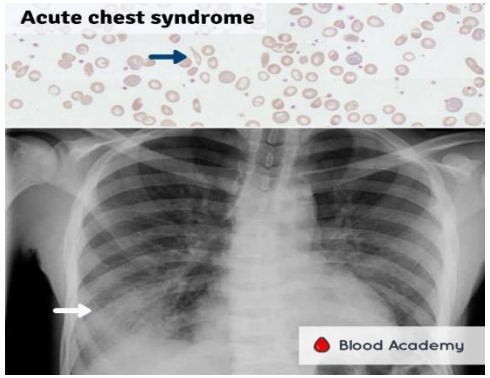
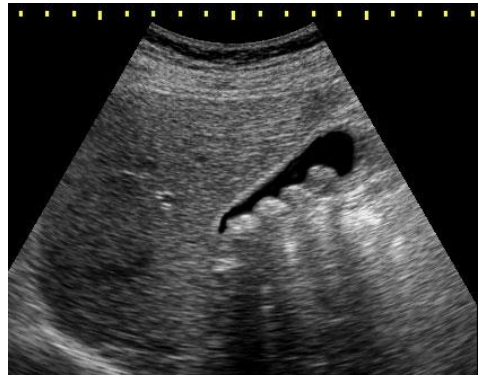
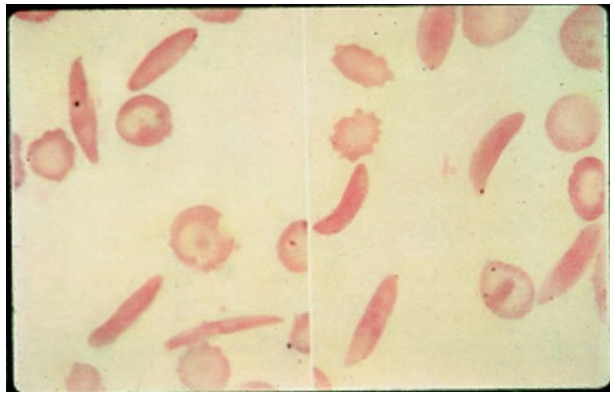
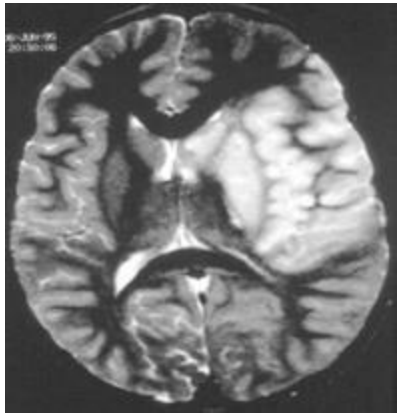
Enuresis







Priapism



How To Manage Pain At Home

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- Distraction techniques – read a book/ play game/meditation
- Drink plenty of fluids
- Keep warm – have a bath/ wheaty bags
- Pain relief:
  - Paracetamol +/- NSAID
  - Opioid medication – codeine/morphine
- If pain not resolved need to attend hospital for consideration of intranasal/ intravenous analgesia/ IV fluids

What can I do to help myself?

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- Try to keep blood flowing freely :
  - Keep warm
  - Keep well hydrated
- General Health:
  - Exercise regularly
  - Eat a good diet with fruit/vegetables for all nutrients
  - Try and manage stress/anxiety
- Take prescribed medicines from the hospital:
  - Penicillin V (to prevent infection)
  - Folic Acid (to give nutrients needed to make red blood cells)
  - Paracetamol/Ibuprofen if pain
- Vaccinations
  - Routine childhood vaccinations and additional flu vaccine/pneumococcal vaccine/Men ACWY
- If unwell need to seek medical advice early

Why Do I Need To Come To Clinic?

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- To provide support with living/caring with sickle cell disease
- To discuss any problems managed at home
- To check bloods and ensure no damage because of sickle cell disease
- To ensure growing/puberty
- To examine to ensure no damage due to sickle cell disease
- To discuss medications available and monitor if taking
- To discuss any new advances in sickle cell disease

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Thank you

Any Questions?

Oluwabukola Akanni
Growing up with SCD

North East & Yorkshire Haemoglobinopathy Coordinating Centre

A Life of Pain, Resilience, and a Plea to Be Heard

“A story of struggle, strength, and a call to action.”

Olu Akanni

What is Sickle Cell Disorder?

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- Sickle Cell Disease (SCD): A genetic condition causing red blood cells to sickle, leading to severe pain, organ damage, and health complications.
- Affects 17,500 people in the UK, millions worldwide.
- Lifelong battle with unpredictable pain crises.

Olu Akanni
A Childhood Caged by Pain

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- Grew up in a loving family in Nigeria, but SCD cast a shadow.
- Siblings enjoyed freedoms (climbing trees, holidays) I couldn’t.
- Parents’ fear confined me to protect my fragile life.
- Missed out on boarding school, longed for stories of the outside world.

Olu Akanni
Nights of Terror

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- Pain crises struck at midnight, bones feeling shattered, body ablaze.
- In Nigeria, nighttime hospital trips were impossible due to danger.
- Family rallied: siblings pressed limbs to dull pain, dad used hot towels.
- Guilt for keeping family awake, longing to escape my body.

Olu Akanni
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Olu Akanni
The Weight of Stigma

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- Friends didn’t understand last-minute cancellations: “You don’t look sick.”
- Mocked as “jellyfish” or “sickler” for slow steps.
- Faith healers claimed I was “possessed” or needed more prayer.
- Hid my condition to avoid judgment, longing to be seen as more than SCD.

Olu Akanni
The Rules That Bound Me

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- Lived by strict rules: no playing hard, no staying out late.
- Envied Serena Williams' strong, pain-free body.
- Each crisis led to promises to never break the rules, but freedom tempted me.
- Parents' fear grew with news of other SCD children dying.

Olu Akanni
Resilience Through Love

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- Family's unwavering love was my lifeline, lifting me when I wanted to give up.
- Earned two degrees and pursued journalism despite SCD's toll.
- Parents' love still guides me, even though they're gone.
- "I'm a warrior, not a 'sickler.'"

Olu Akanni
A New Hope – Peer Mentoring

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- Now a peer mentor with the Sickle Cell Society, supporting youth aged 10–24.
- Helping others build self-worth and connect, reducing SCD's isolation.
- "My childhood was lonely; I'm ensuring others don't face that."

Olu Akanni
Be Our Champions

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- SCD warriors are resilient, hardworking, and independent, but we need support.
- Advancements help us live longer, but challenges remain: chronic pain, need for better care.
- Call to action: “Tell It Loud” – learn about SCD, advocate for better care, support sicklecellsociety.org.
- Thank you to existing champions (parents, carers, healthcare workers).

Olu Akanni
Help Us Live

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- “We want to live, help us to live.”
- SCD taught me challenges don’t define you—community does.
- Let’s amplify SCD voices, reduce stigma, and build a world where we thrive.
- Thank you for listening—let’s keep telling our stories loud and proud.

Your Diet Your Health:
Considering Healthy Eating Principles, Habits and Behaviours

Presented by

Dr Claudine Matthews DPROF HSC RD FHEA

NE & Y Patient Education Event - Nutrition

21st June 2025

Education Session (2) Agenda:

Part 1: Why good nutrition is important: Be intentional with your diet and lifestyle choices

Part 2: The Sickle Cell Nutrition Eatwell guide – healthy principles to eat and live with

Part 3: Empowerment to improve your quality-of-life outcomes

Part One:

1. Why Good Nutrition is Important:

Be intentional with your diet and lifestyle choices

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
  - Growth Delays
  - Delayed Puberty
  - Impaired immunity
  - Increased risk of stroke
- Management
  - Eating a Healthy Balanced Diet
  - Engage in healthy lifestyle activities like regular activity- BUT
  - Don’t smoke, limit alcohol intake

Why is good nutrition important?

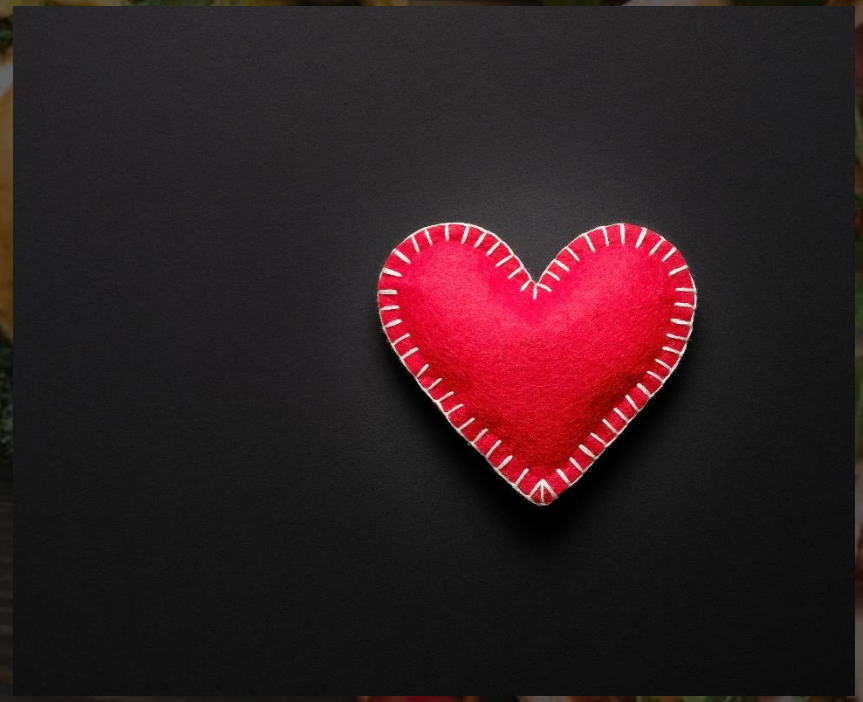
Building a good nutritional status and **preventing Malnutrition:** → A **good nutritional status** is all about you -

Eating a healthy balanced diet, Healthy weight → Eating the **right type and amounts of food**

Choosing and eating the **right variety of food** → Have all the nutrients your body needs to enjoy a **good quality of LIFE**

Healthy Eating habits and Behaviours

- Your Diet Your Health
  - We all have a CHOICE to make
  - Will it be Chicken and chips, KFC or MD's or a HEALTHY Balanced DIET
  - Knowledge is **POWER**
  - Knowledge without **ACTION** is **INACTION**
  - **START EARLY to** build a **FOUNDATION** for **long term HEALTH** and **WELLBEING**
  - WHAT are you going to CHOOSE…??



Part Two:

2. Sickle Cell Nutrition Eatwell Guide – healthy principles to Live well with sickle cell

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.

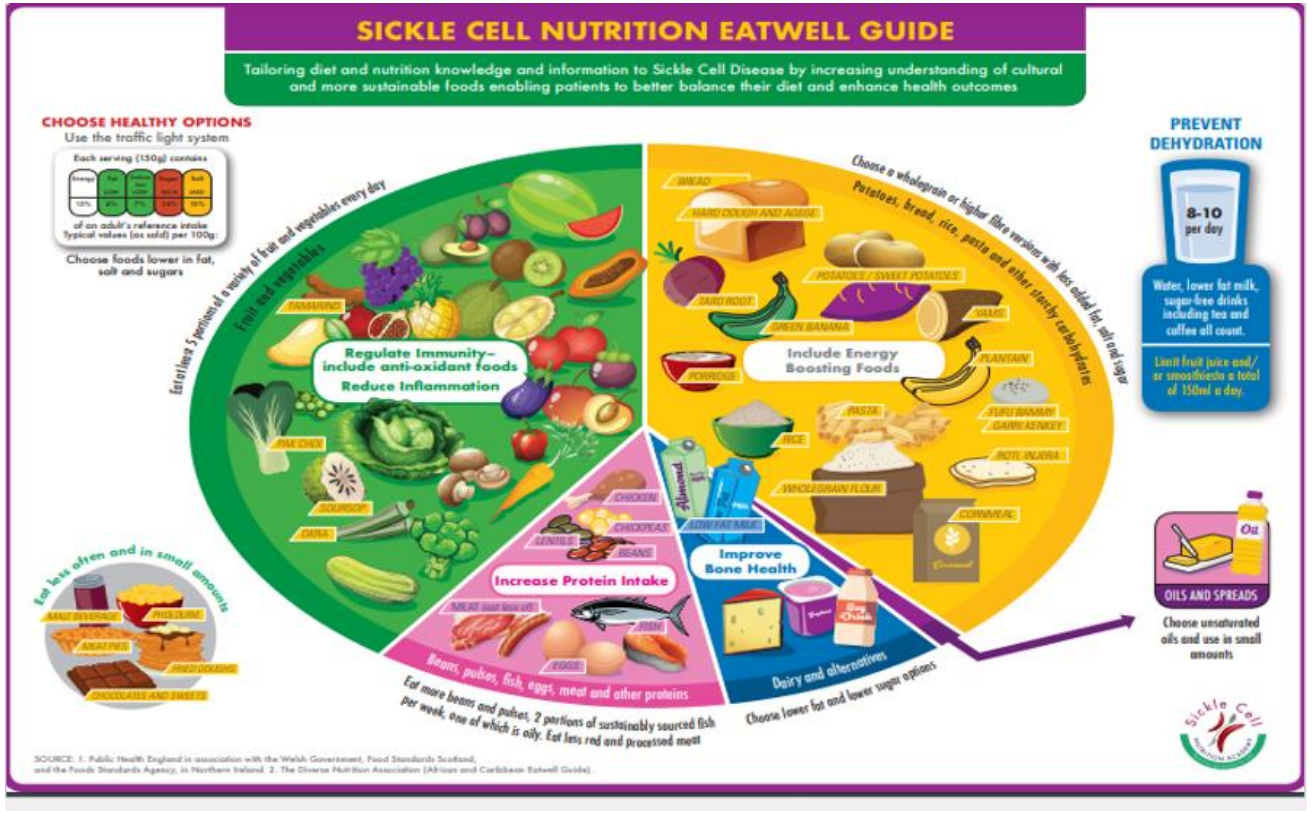
OILS AND SPREADS

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).

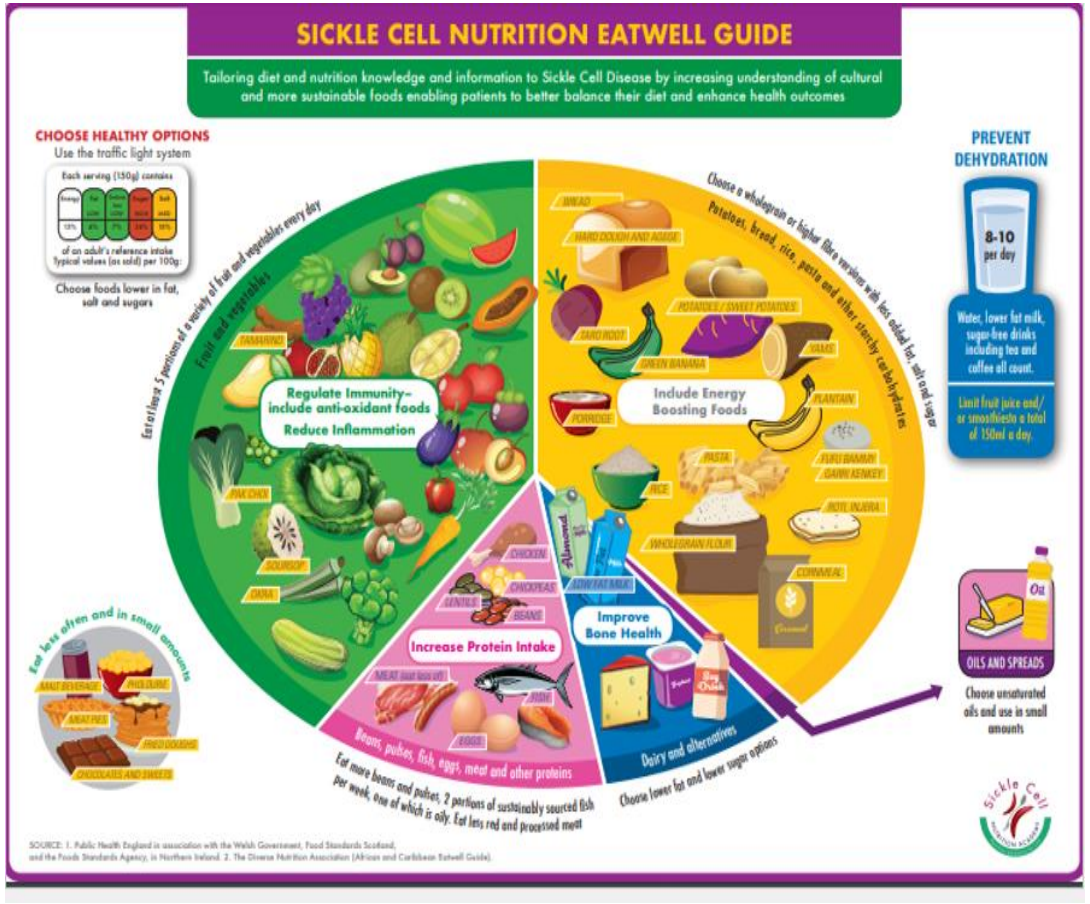
7 Key Healthy Messages in the SCD Eatwell Guide

- 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

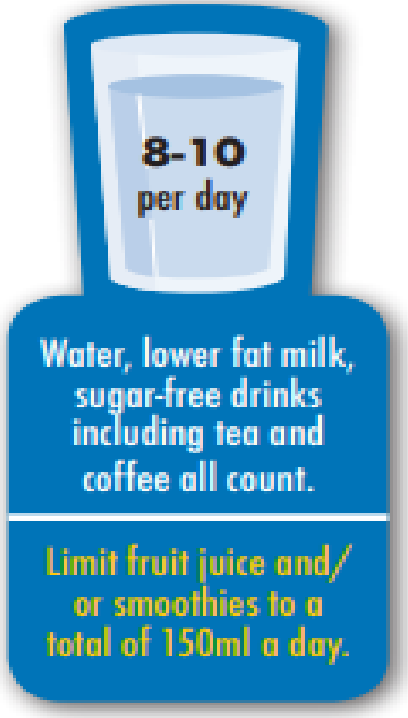
- Carbohydrates (CHO)- breads, cereals, potatoes, rice, pasta, yam, plantain
- Fruit and Vegetables (Your 5 a Day)
- Proteins - Beans, pulses, meats and other protein, fish, eggs, other proteins
- Dairy, milk and milk alternatives, cheese, yogurt
- Fats and oils



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

Each serving (150g) contains

Energy	Fat	Saturation	Sugar	Salt
	LOW	LOW	HIGH	MED
13%	4%	7%	36%	15%

of an adult's reference intake
Typical values (as sold) per 100g:

Choose foods lower in fat, salt and sugars

Sickle Cell Nutrition Academy

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 Breakfast Ideas

Healthy Lunch Ideas

Healthy Dinner options

Healthy Traditional Dinner Options

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

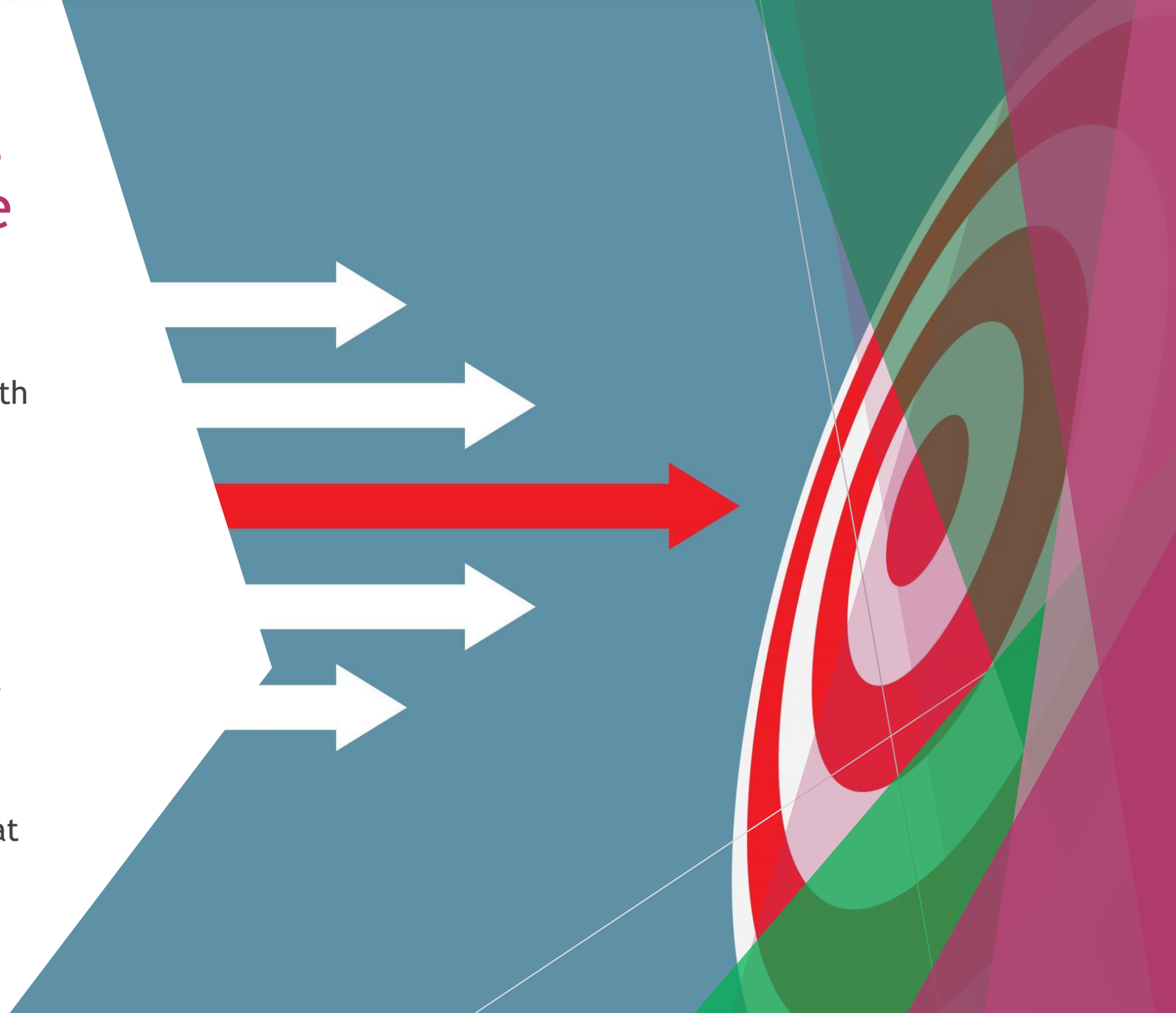
Healthy Snack Options

Part Three:

3.Empowerment to improve your quality-of-life outcomes

Factors influencing your Quality of Life

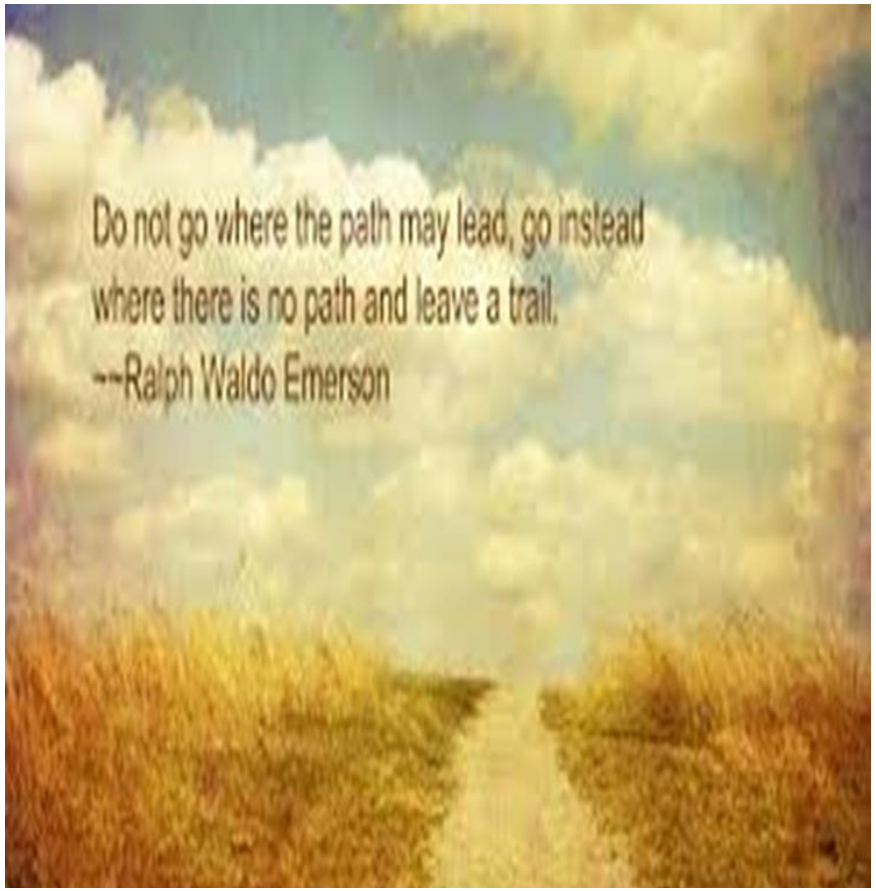
- **Goals (Outcomes)**
- Have you set goals for your health and wellbeing?
- **Expectations (Hopes)**
- What are your expectations of your health and what is influencing it?
- **Standards (Excellence)**
- What standards have you set for yourself and your Health?
- **Concerns (Worries)**
- What are your concerns and what help do you need managing it?



One thing you will

- Take Away…

ONE TAKE AWAY…

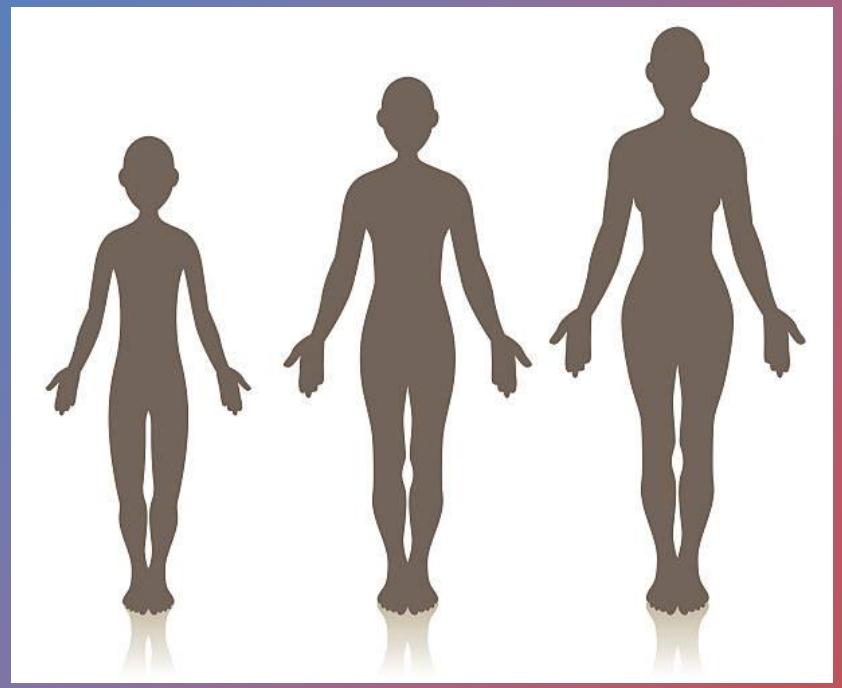


Thank You!

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

Growing Up with Sickle Cell

What Every Girl Should Know



What is Puberty?

- Natural body changes to adulthood
- Breast development, hair growth, periods (menstruation)
- Growth spurts, mood swings

When Should Puberty Start?

- Usually between 8 and 13 years
- Can be later in sickle cell
- If no breast development by 14 or no periods by 16 - talk to your nurse or doctor
- Everyone develops at their own pace

Sore Breasts

- Breasts can be uncomfortable when first developing
- Breast tenderness also common before periods
- Helped by well-fitting bras
- Consider getting measured in a bra shop (or at home)
- Try padded bras if you have tenderness

Periods – they can be a Pain!

- Cramps
- Heavy bleeding – see your doctor if a sanitary pad or tampon lasts less than 2 hours
- Feeling tired

Periods can cause sickle pain

- Some people have more sickle cell pain around their periods
- If this happens to you, tell your nurse or doctor
- Extra medication can help

Looking after Yourself around Periods

- Stay hydrated (not too much caffeine – or in the future alcohol)
- Eat a healthy diet
- Rest and relax
- Take pain relief, hot water bottles
- Talk to someone about how you feel
- Hormonal medication might help if periods painful or heavy, or bring on crisis

Mental and Emotional Health

- Puberty brings mood changes
- Periods affect your mood as well
- Sickle cell can cause stress
- You are not alone – talk to friends, family, nurses, doctors, psychologists, mentors
- Support groups can help
- Join the Sickle Cell Society

SICKLE CELL SOCIETY

Fertility and Sexual Health

- Most women with sickle cell can have children
- Talk to your healthcare team first if you are planning to have a baby
- Use contraception unless you are at a stage in life when you are trying to get pregnant

-condoms, pill, implant, injection, patch, coil etc

- Only condoms protect against STIs
- ‘Natural methods’ of contraception are not reliable!
- Best to plan a pregnancy and talk to your doctor first

Conclusion

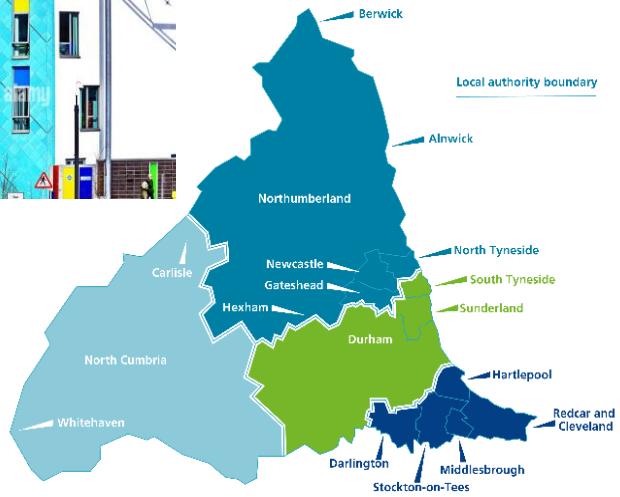
- Puberty with sickle cell can be different, everyone is unique
- If puberty or periods are difficult – talk to someone, we can help!
- Ask questions and take care of yourself ☺

Questions?

Speaker Bio

North East & Yorkshire Haemoglobinopathy Coordinating Centre

Jenni Lawrence
Children & Young People's
Haemoglobinopathy Clinical Nurse Specialist
Great North Children's Hospital
Newcastle upon Tyne



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Medication in Sickle Cell Disease

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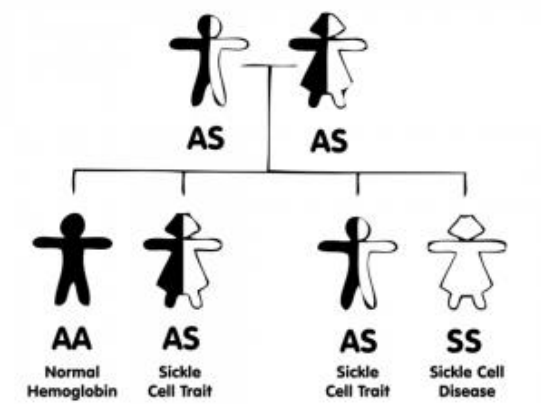
What is Sickle Cell Disease?

- Inherited condition
- Affects red blood cells
- Haemoglobin S
- Red cells change shape, get hard and sticky and block blood vessels
- Red cells break more easily
- Leads to pain and anaemia



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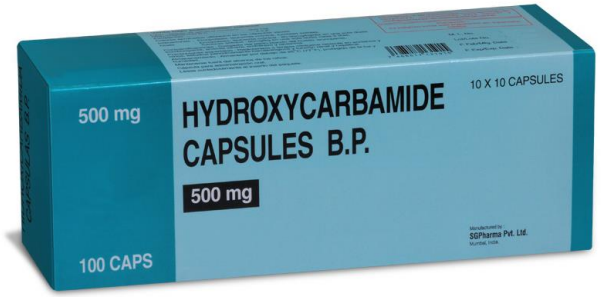
Medicines for Sickle Cell Disease



- Treatment for Sickle Cell
- Makes blood less sticky
- Helps stop crisis

- Increases HbF (Reduces HbS)
- Reduces inflammation
- Works over a long time
- Few side effects

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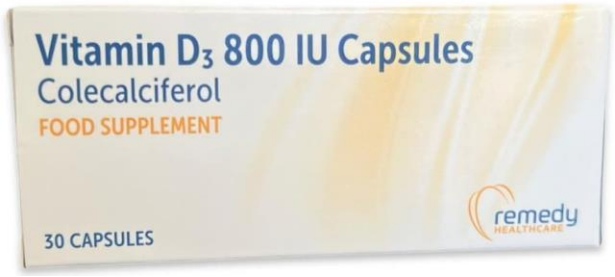
Medicines for Sickle Cell Disease

- Antibiotic
- Taken twice a day
- Helps protect against infection

- Vitamin
- Can be found in food
- Helps to make blood cells



- Vitamin
- Can be found in food
- Helps keep bones healthy
- Helps support immune system

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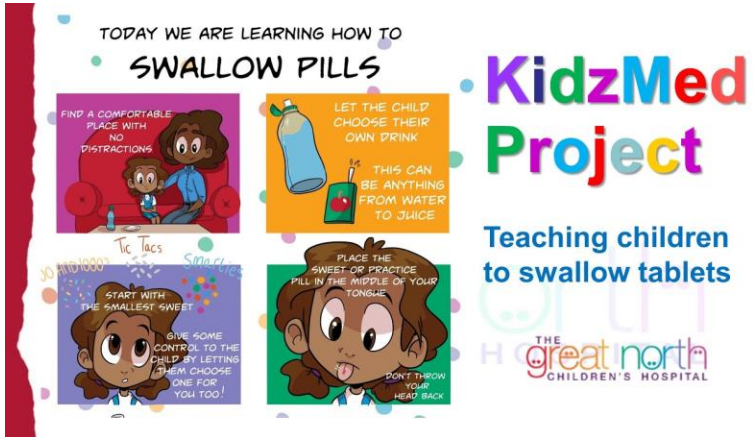
Switching to tablets



Switching to tablets from liquid medication is good for lots of reasons:

- Easier to carry
- Doesn't need to go in the fridge
- Longer shelf life
- Some liquids don't taste nice
- **Better for teeth**

KidzMed can help – ask your hospital team!

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How can I remember to take my medicine?

Work out what works for you!

- Keep medicines somewhere safe but sensible
- Coordinate with another activity
- Set alarms
- Pill boxes can help you stay on track

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What else can I do?

- Medicine is very important, but there are some other things that can help:
  - Hydration
  - Warmth
  - Relaxation
  - Gentle exercise

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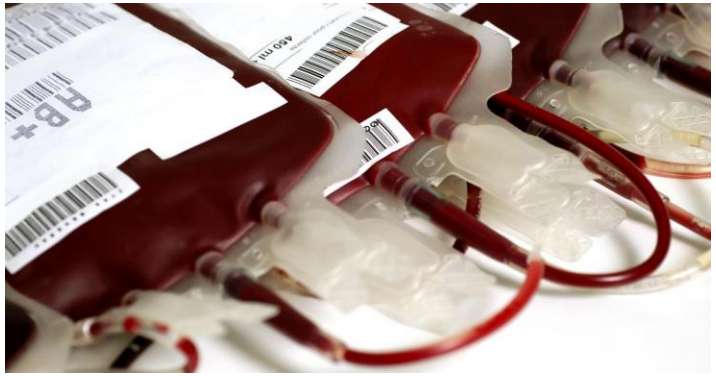
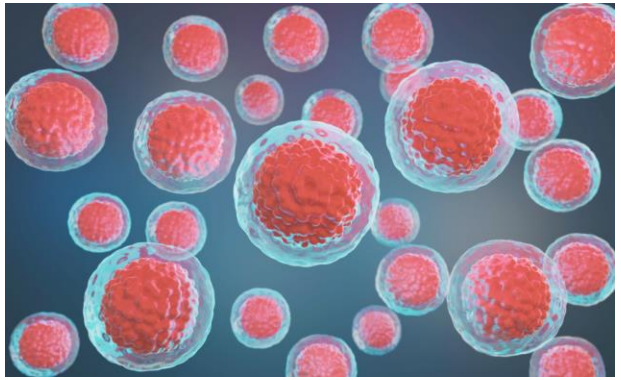
Are there any other treatments?

Red Cell Exchange



Blood Transfusions



Stem Cell Transplant/
Gene Therapy

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Thank you

Cheryl Hall, Polly Crookes, Carly Bell

Haematology Nurse Specialists, Sheffield Children's hospital

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Transition

What is Transition and why do we do it?

What exactly is transition?

- Transition is the process of moving your Sickle Cell Care from Your Children's Hospital to your Adult Hospital.
- The process starts at around 14yrs.But can start earlier.
- You will be invited to additional Transition clinics alongside your usual appointments.

Why do we use the Transition Process

01

To prepare young people for adult services.

02

To increase your knowledge around Sickle Cell and the care you receive

03

To increase your confidence

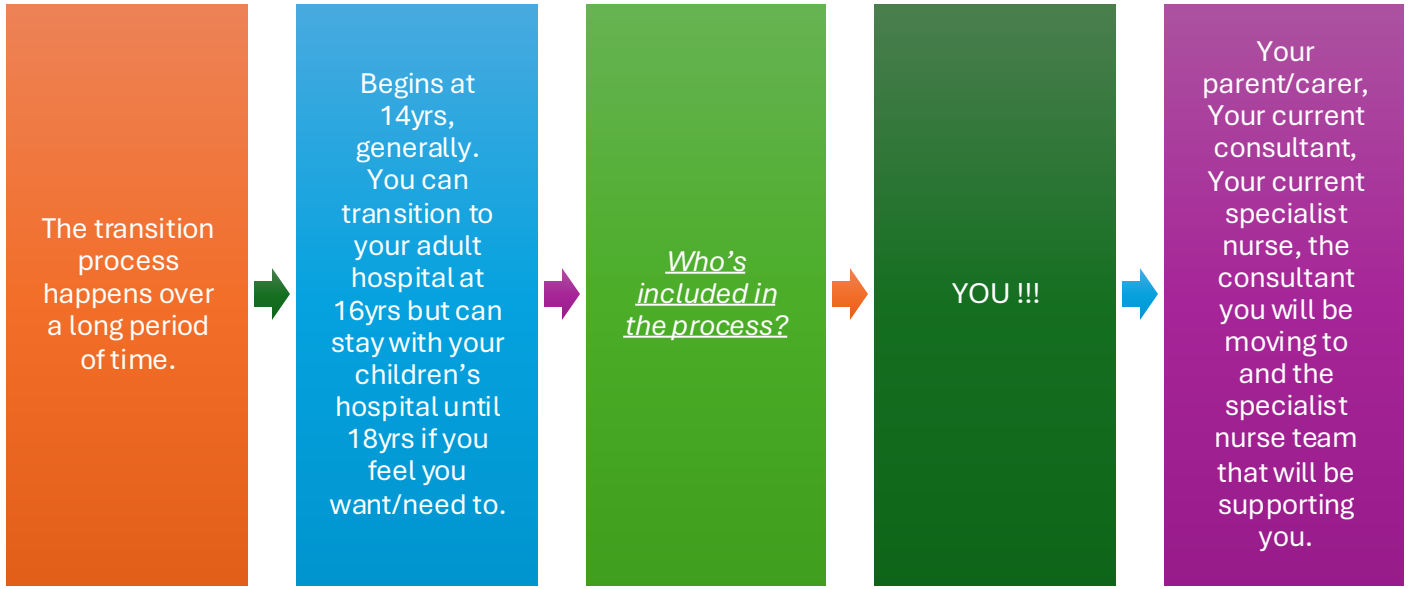
04

To increase your independence

05

To support you in managing your health

Transition process -



Transition is important because -

Teenagers and young adults are massive risk takers-

May not take medications at all or irregularly

May put themselves in high-risk situations

Outside influences can affect compliance with medications and self care

Brain Development isn't complete –

Although you may think it is, your brain isn't fully developed until about 25yrs.

Outside influences and other people's opinions, idea's, cultural Norms may influence how you feel about your health and how you care for yourself.

Massive time of change – Puberty, Exams, leaving home to go to university etc

Transition is important because -

- All of these factors have a massive impact on how you feel about your health, how you treat your own health especially Your Sickle Cell health, and how you look after your health and well-being in general.

The aim of transition

To move from paediatrics

Parents /Carers giving and managing medicines

Parents/carers arranging appointments

Parents/carers Talking to Dr's & Nurses

Parents/carers making all the decisions

To the adult Hospital

YOU taking medications independently

YOU knowing about your Sickle Cell and how to manage it

YOU arranging your hospital appointments

YOU talking to the Dr's and Nurses about your care

Tools used to help the transition process

- Transition Clinics – Generally done in your children's hospital, Adult Consultant and Nurses attend to discuss with you how they work and what to expect.
- Ready steady go – Questionnaire style papers that aim to find out your knowledge and understanding of Sickle Cell and medications.
- Ask 3 Questions – 3 questions that you can ask your Dr /Nurse to help you understand any questions regarding your sickle Cell care.
- Health Passport – Information regarding your health that you can use to help any new Dr's /Nurses you meet understand how you manage your Sickle Cell.

Any Questions ?