Bitesize sessions

WHAT ARE SICKLE CELL DISORDERS?

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DISCLAIMER: These slides are for education purposes only.
Speak to your healthcare professional if you have any questions relating to your health.

I WILL COVER...

- All about red cells
- The background of sickle cell disorders
- How sickle cells affect the body (complications)
- Helping to prevent sickle-related complications in the workplace

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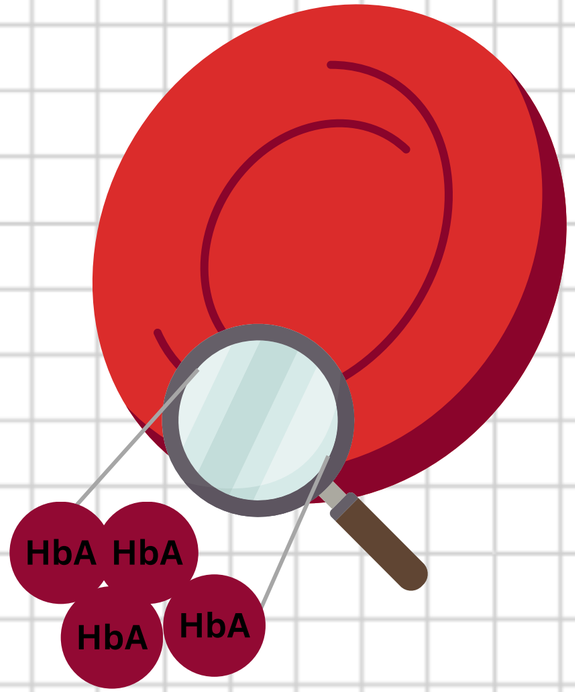
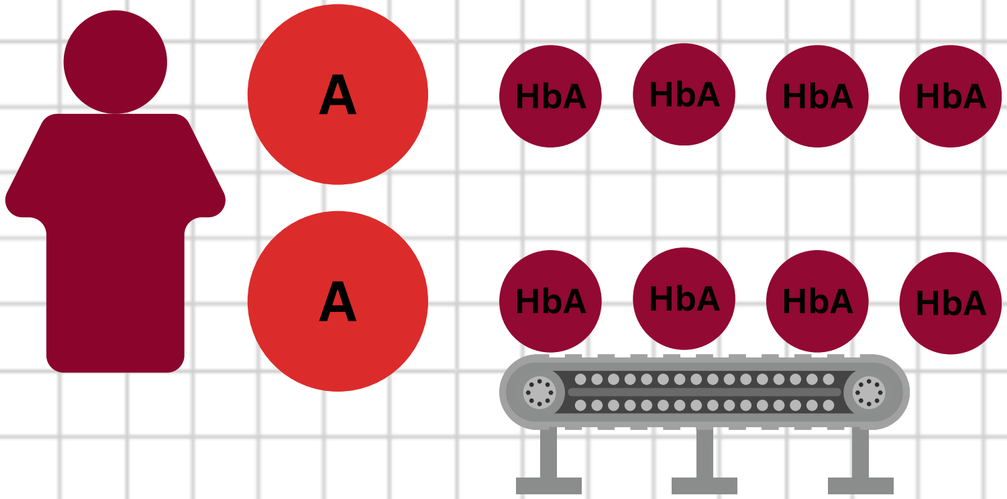
RED CELLS

- Red cells are blood cells which travel around the body in the blood stream
- They are filled with proteins called **haemoglobin** which carry the oxygen
- Haemoglobin is made by the two genes which are inherited - one from each parent
- Most people have the same kind of haemoglobin - Haemoglobin A, also known as **HbA**.





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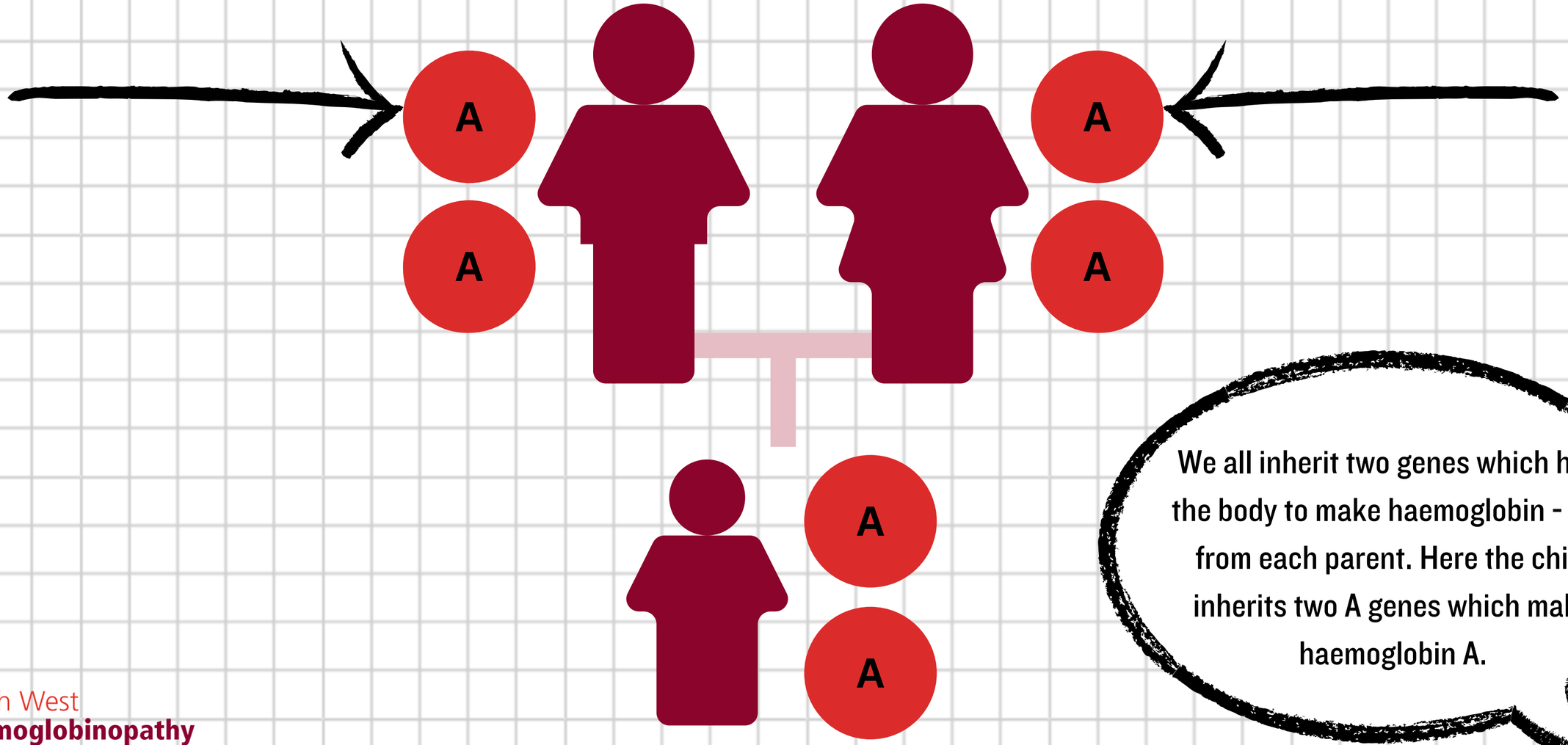
GENES



We all inherit two genes which help the body to make haemoglobin - one from each parent. Here the child inherits two A genes which make haemoglobin A.

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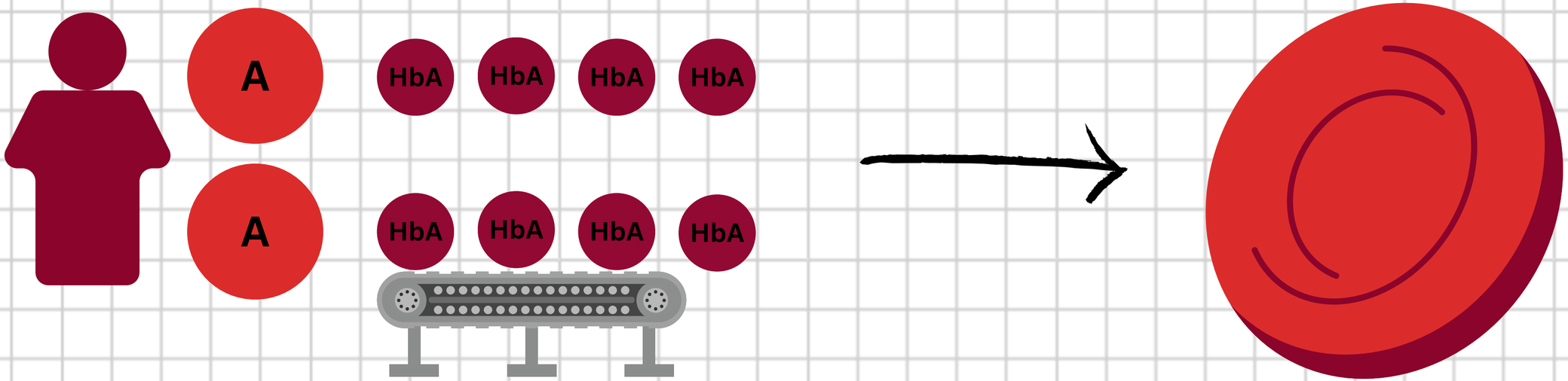
The two A genes allow the bone marrow (the blood factory of the body) to make a constant, free-flowing supply of haemoglobin A which forms healthy, disc shaped red cells.



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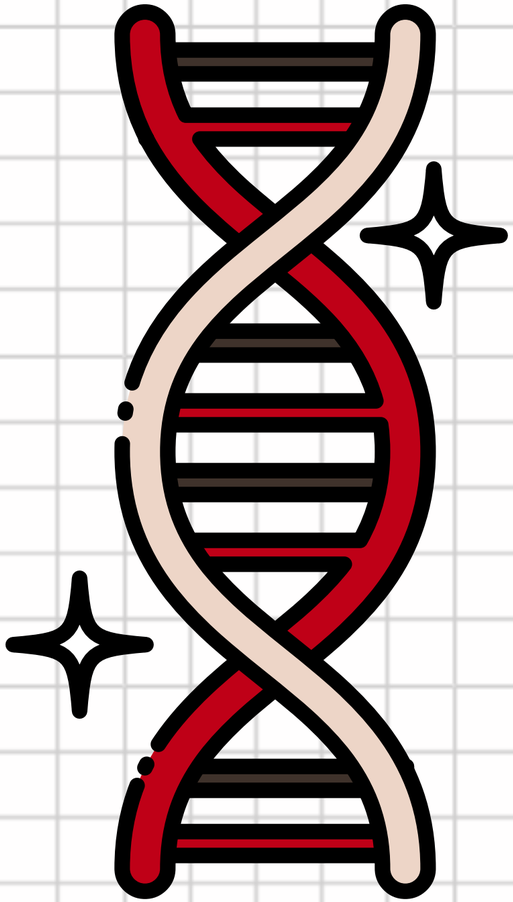
What happens when the haemoglobin genes are abnormal?



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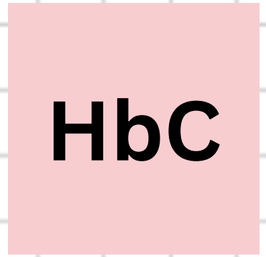
HAEMOGLOBINOPATHIES

- Sometimes the genes which create haemoglobin have a fault in their coding.
- This leads to the creation of misshapen haemoglobin - this is called a **haemoglobinopathy**
- The different types of haemoglobin have different names, for example **HbS**, **HbC**, **HbD**.... etc. There are more than 1,000 different types of haemoglobin in existance.
- These conditions are **inherited.**



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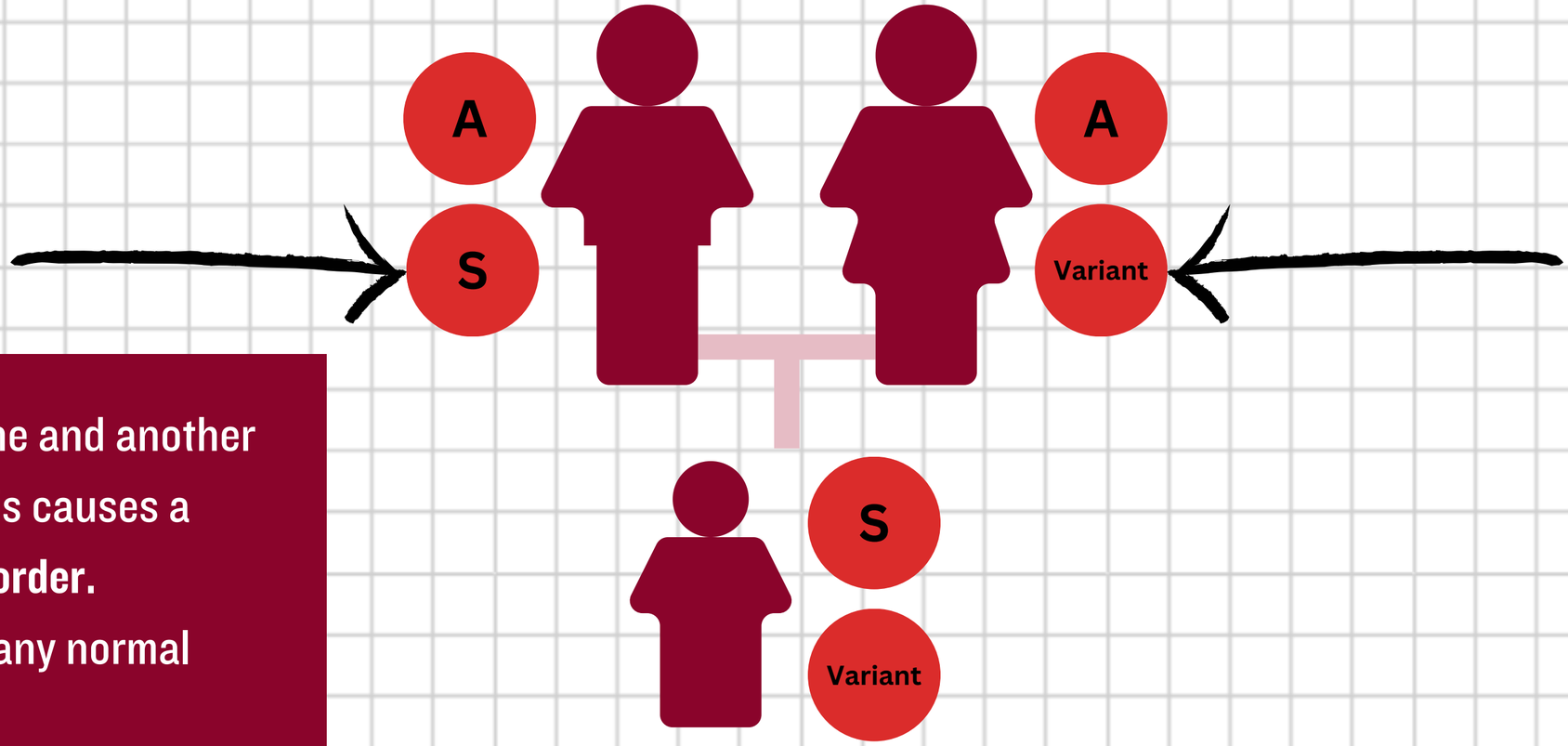
SICKLE CELL DISORDERS



- When a person inherits an S gene and another abnormal haemoglobin gene, this causes a condition called a **sickle cell disorder.**
- These individuals cannot make any normal haemoglobin.

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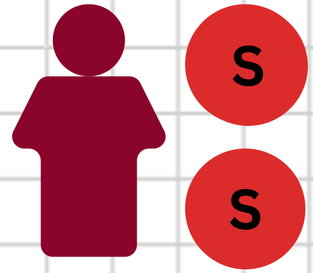
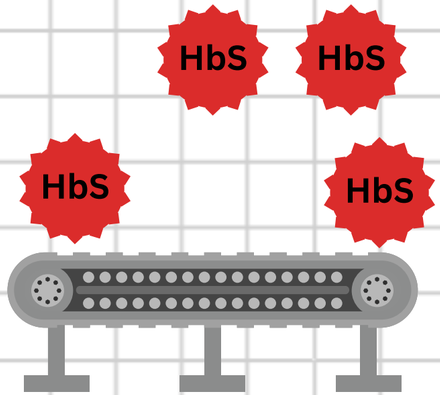
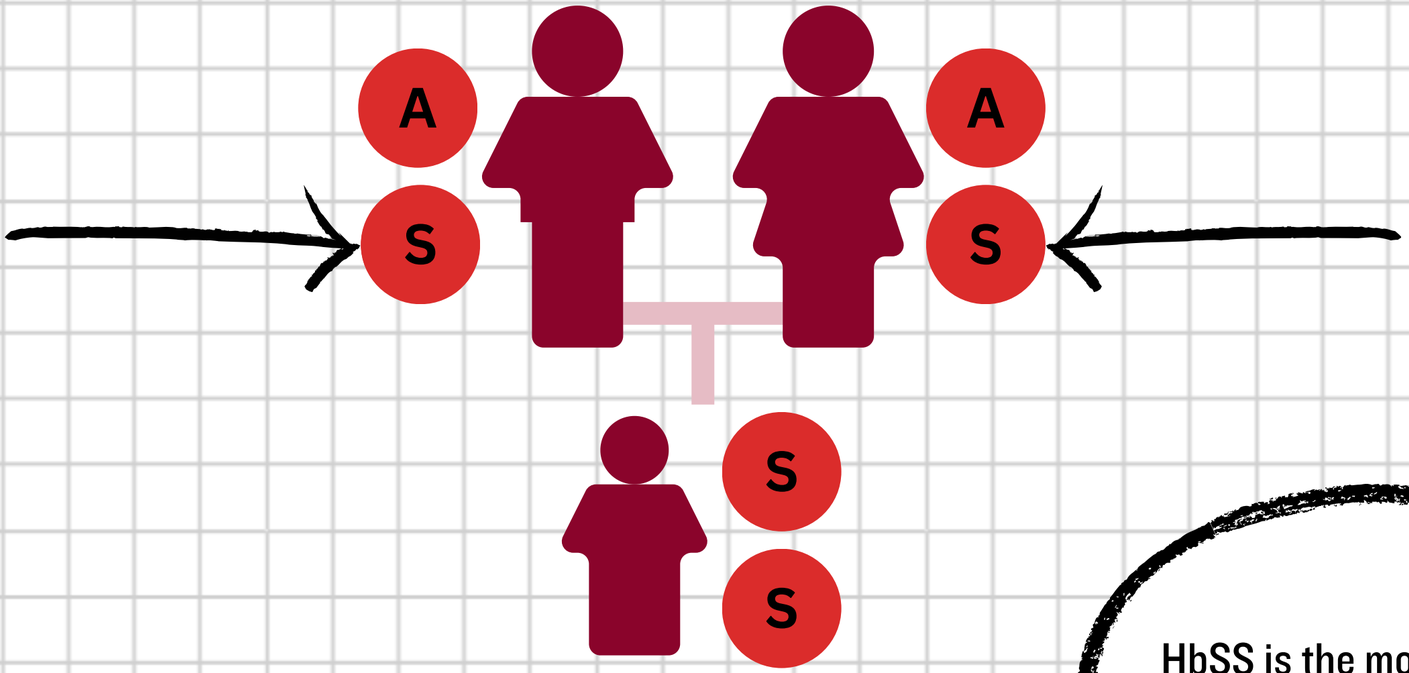
SICKLE CELL – HbSS

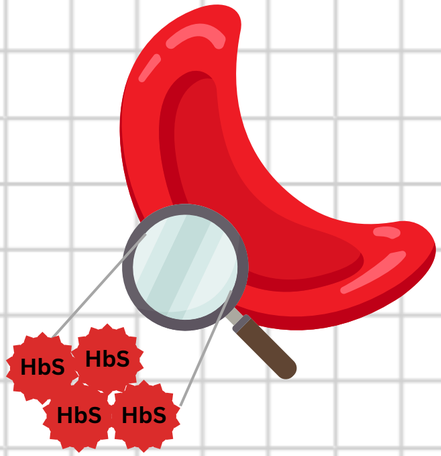
The production is also slower than normal when the haemoglobin gene is faulty.







HbSS is the most common type of sickle cell disorder.



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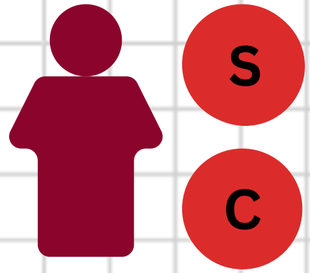
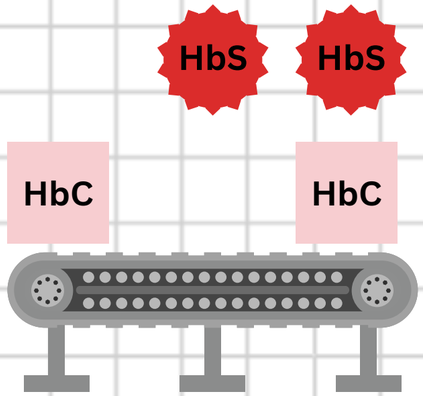
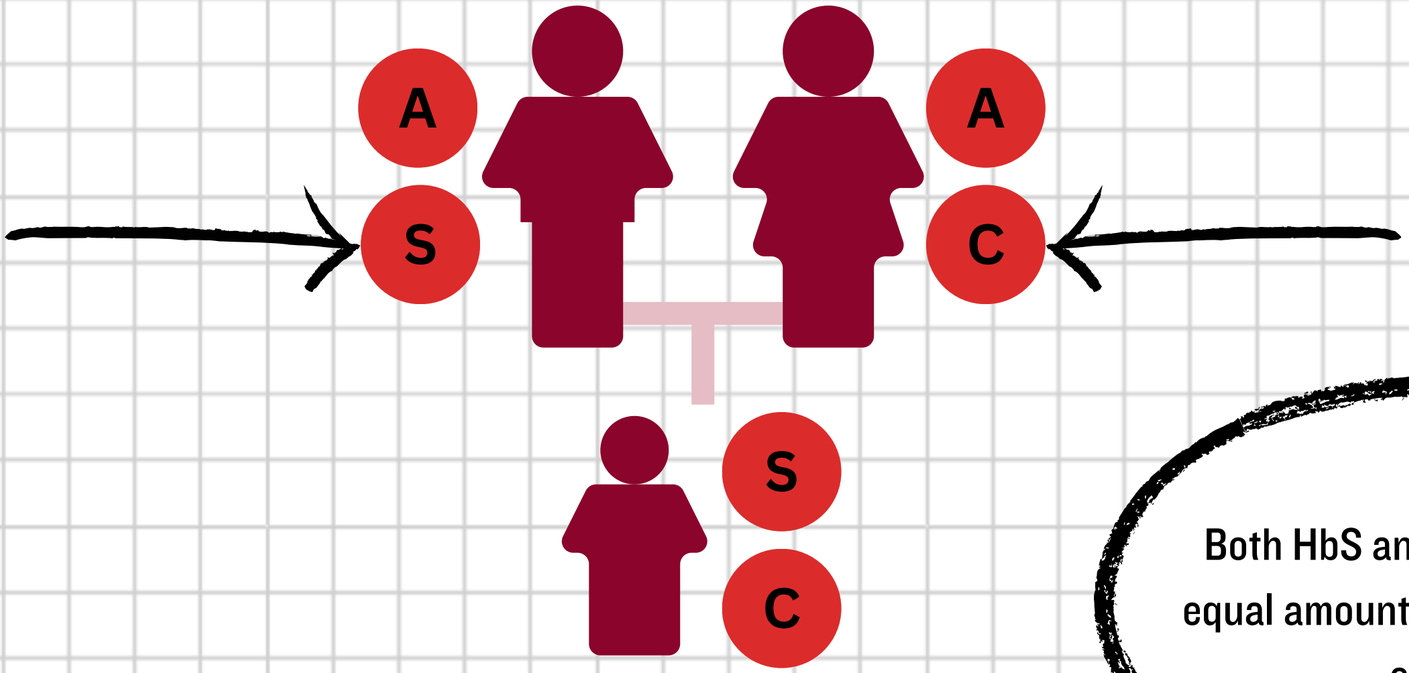
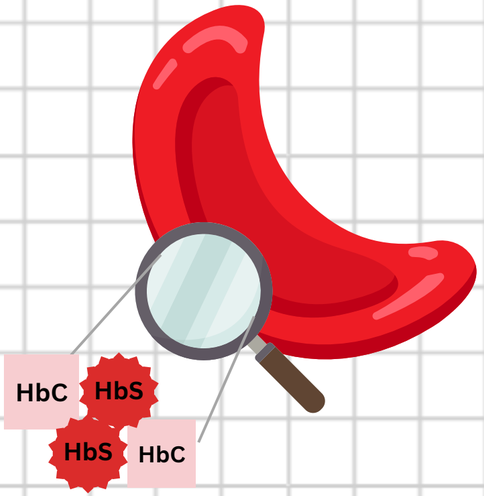
SICKLE CELL – HbSC







Both HbS and HbC exist in equal amounts inside the red cell.



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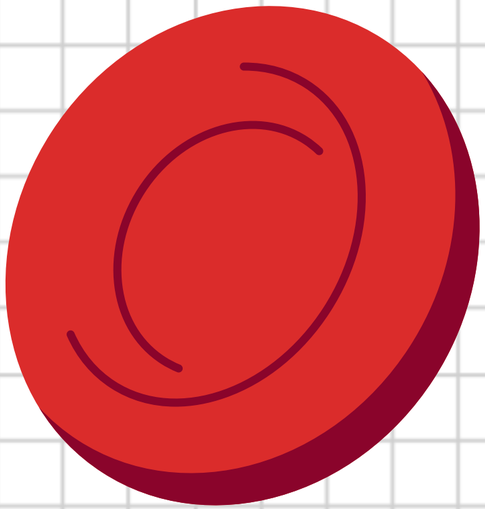
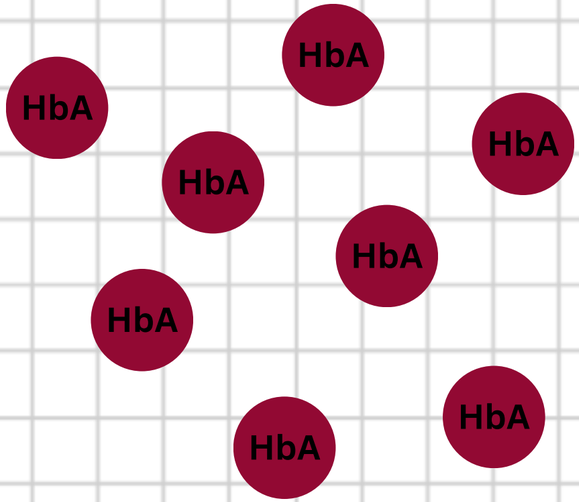
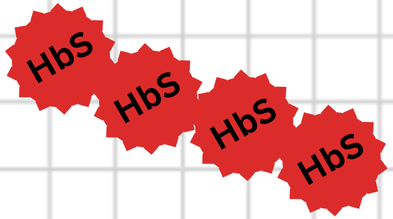
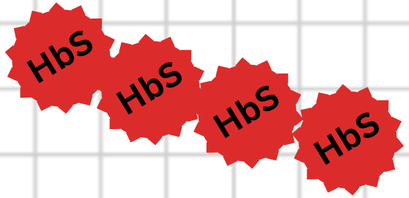
SICKLE CELL DISORDERS

WHY DO CELLS SICKLE?









The abnormal haemoglobins easily stick together inside the red cell, particularly in times of stress, dehydration and exertion. This causes the cells to form the sickle shape.

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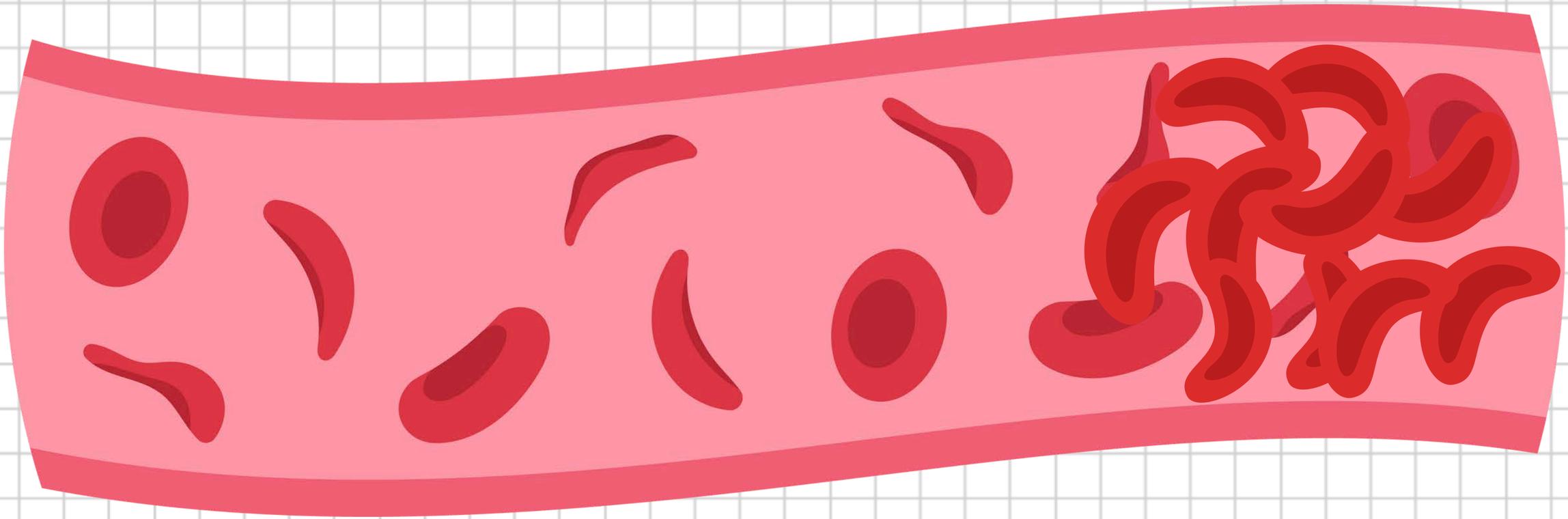
WHAT DO SICKLED CELLS DO IN THE BODY?



Under stress, red cells turn into a sickle shape. This blocks small blood vessels, blocking off the blood supply to the area and causing **extreme pain.**

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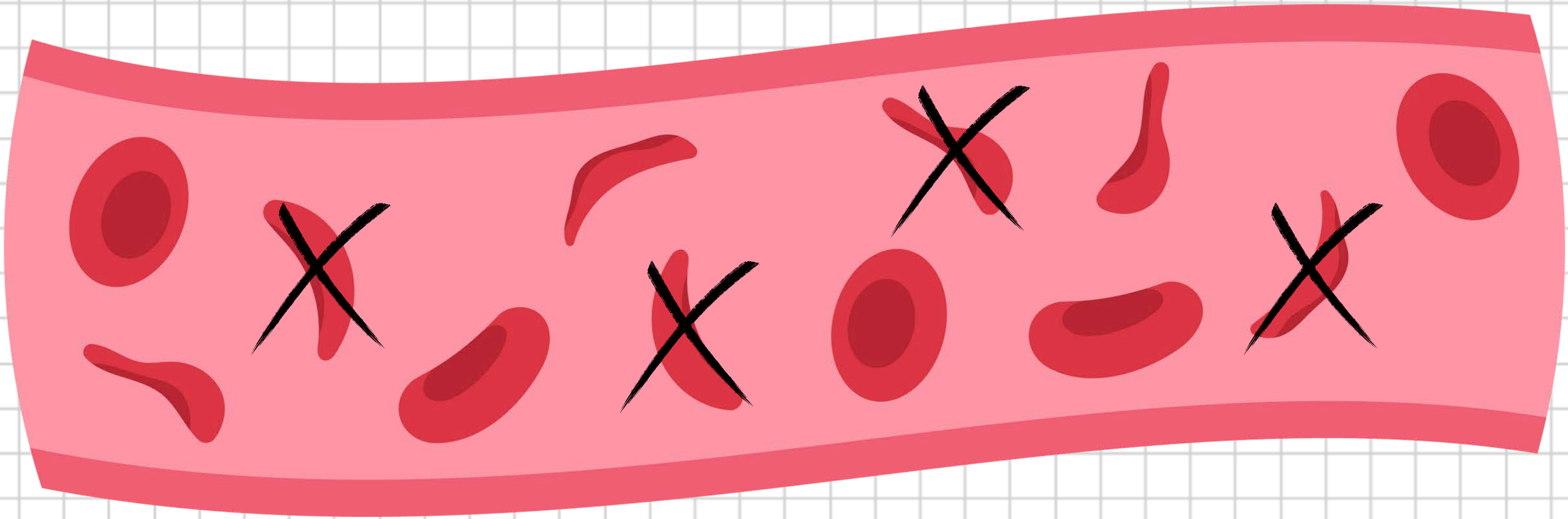
WHAT DO SICKLED CELLS DO IN THE BODY?



The body recognises the sickle shaped cells as abnormal and destroys them. This causes a worsening **anaemia**. This can contribute to fatigue.

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COMPLICATIONS OF SICKLE CELL DISORDERS

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URGENT COMPLICATION

PAINFUL CRISIS

- Sickle cells can block the blood supply to any part of the body, but is common in the bones
- When the bone loses it’s blood supply, this causes extreme pain
- Triggers for painful crisis can include:
  - Cold weather
  - Infection
  - Dehydration

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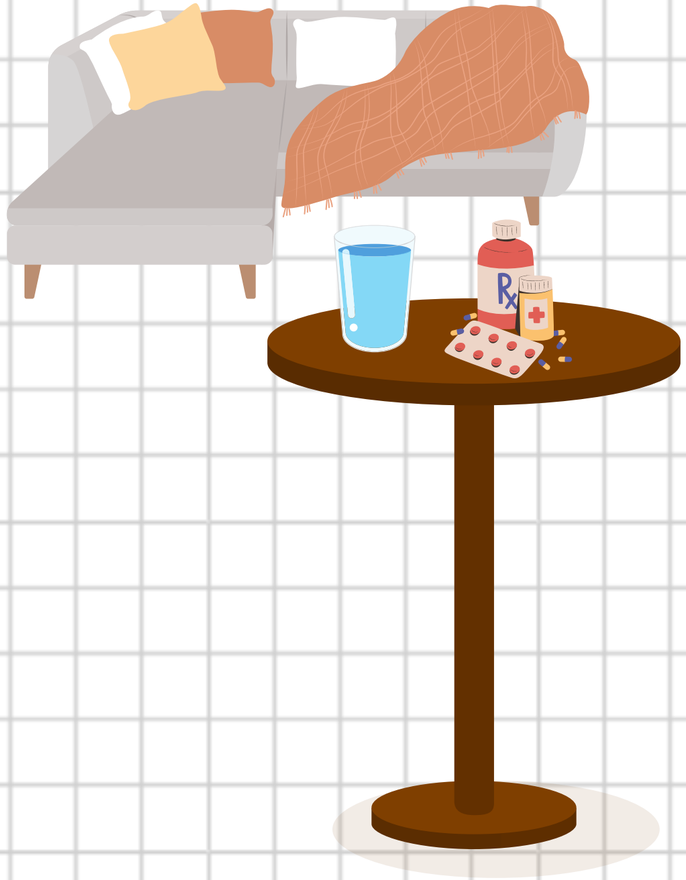
URGENT COMPLICATION

HOW IS A PAINFUL CRISIS MANAGED?

- Patients will often try to manage at home at first, with:
  - Rest and keeping warm
  - Regular painkillers
  - Keeping well hydrated
- If the pain continues despite these steps:
  - The patient will attend hospital
  - They often require strong painkillers (morphine) under the skin
  - May be admitted to the ward for monitoring.



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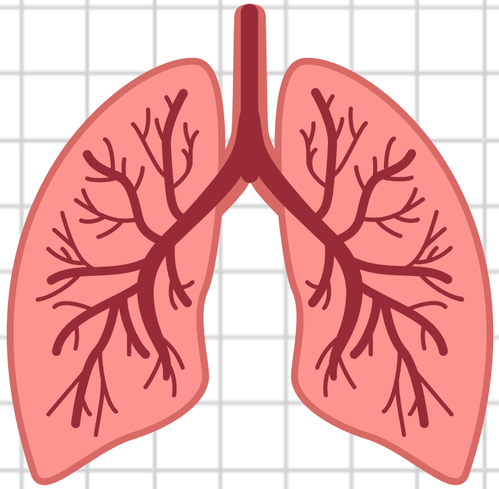
URGENT COMPLICATION



ACUTE CHEST SYNDROME

- A serious complication called **acute chest syndrome** can develop during a painful crisis
- This is where sickle cells block the blood supply to the lung, causing pain and difficulty breathing
- This can be life threatening if not recognised and managed promptly.

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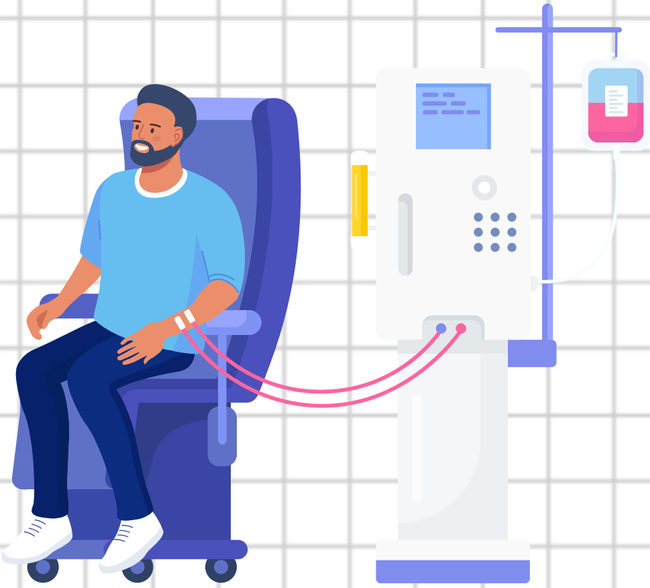
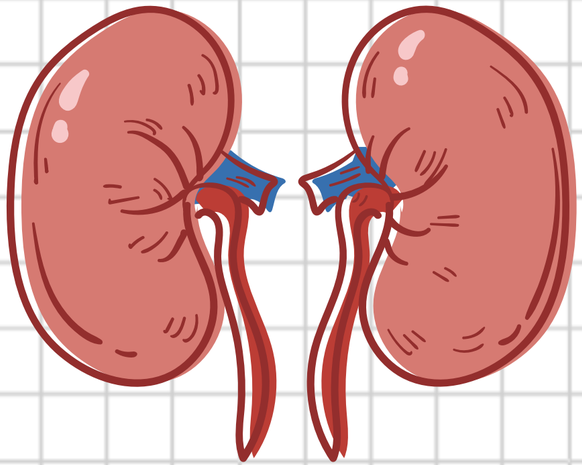
URGENT COMPLICATION

STROKE

- Sickle cells can block small vessels inside the brain
- This blockage can cause damage to the brain tissue, leading to a stroke.
    - Symptoms may include arm/leg weakness, facial droop and difficulty speaking
- A patient who thinks they are having a stroke needs to attend A&E immediately. They would require a life-saving procedure called a red cell exchange, where sickle cells are removed by a specialist machine and healthy cells given back.



LONG-TERM COMPLICATION



KIDNEY DAMAGE

- Over time, sickle cells can cause background damage to the kidneys
- Because of this, all patients with sickle cell disorders pass dilute urine as their kidneys are not able to hold onto the water effectively.
    - Patients with sickle cell disorders must keep hydrated to prevent dehydration, which can lead to painful crisis
    - They may need to use the bathroom more than normal as the kidneys make a lot of urine.

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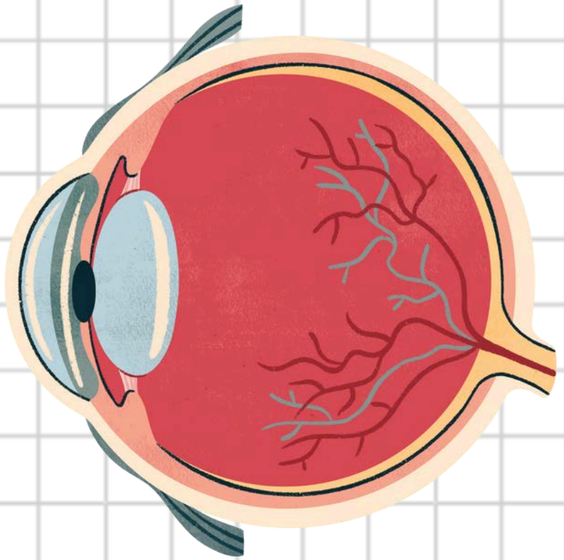
LONG-TERM COMPLICATION

RETINOPATHY (EYE DAMAGE)

- Sickle cells block vessels at the back of the eye, leading to background eye damage at the back of the eye (retina)
- HbSC is more likely to affect the eye
- This can affect the vision including:
  - Floaters
  - Blurred vision
  - Flashing lights
- This is monitored once a year in the eye clinic.



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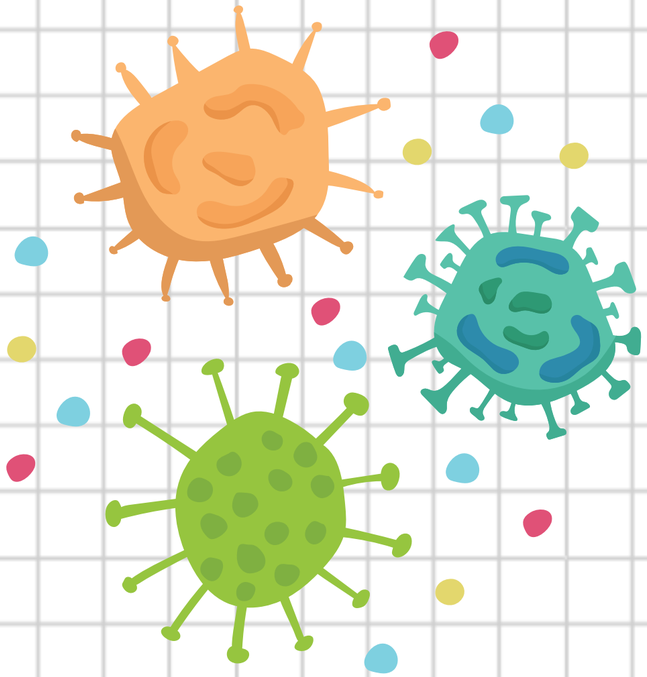
LONG-TERM COMPLICATION



INFECTION RISK

- Patients with sickle cell disorders are at a higher infection risk as their spleen does not work as normal.
- The spleen is an organ which sits under the ribs on the left side. This usually fights certain types of bacteria (called encapsulated bacteria). These bacteria cause infections such as meningitis and pneumonia
- To combat this, patients take lifelong penicillin and must keep up to date with extra vaccinations.

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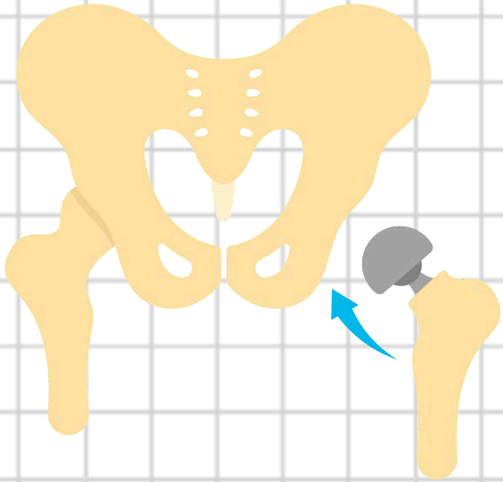
LONG-TERM COMPLICATION

AVASCULAR NECROSIS OF THE HIP

- Sickle cells can cause damage to the hip bone over time
- This can cause issues including:
  - Clicking of the hip
  - Chronic pain
  - Difficulty walking, requiring walking aids
- Many patients require a hip replacement to improve the symptoms.



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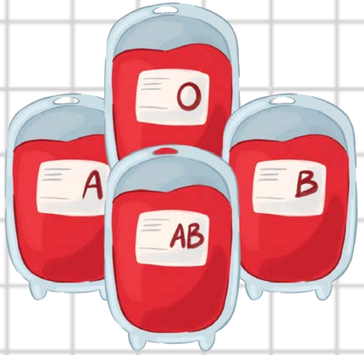
SICKLE CELL DISORDERS

WHAT TREATMENTS ARE AVAILABLE?

Hydroxycarbamide

- A tablet which increases the amount of baby (fetal) haemoglobin - this prevents the cell from sickling as easily
- Taken daily
- Side effects include nail damage and low blood counts



Blood transfusion or red cell exchange

- Blood transfusions can be given to patients who have had serious complications to prevent them happening again

Transplant and gene therapy

- Stem cell transplant and gene therapy are up-and-coming, potentially curative treatments

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SICKLE CELL DISORDERS

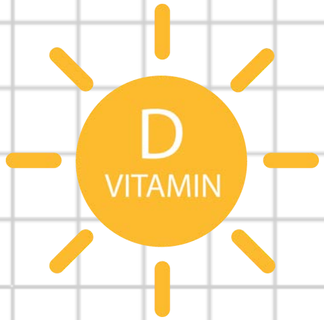
SUPPORTIVE MEDICATIONS

Penicillin

- Helps to protect against infections such as meningitis and pneumonia as the spleen does not work properly
- Taken twice a day lifelong

Folic acid

- An important vitamin which helps to make red blood cells
- Helps to support the body in replacing the red cells which are broken down
- Taken once daily lifelong



Vitamin D tablets

- Vitamin D is important to keep the bones healthy
- Extra vitamin D is needed for patients with SCD to support their bone health
- Taken once daily lifelong

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SICKLE CELL DISORDERS

HOW DO PEOPLE WITH SCD LOOK AFTER THEMSELVES?

Rest when feeling run down or unwell

Keep well hydrated at all times

Keep warm and avoid cold environments

Attend hospital appointments, including blood tests and scans

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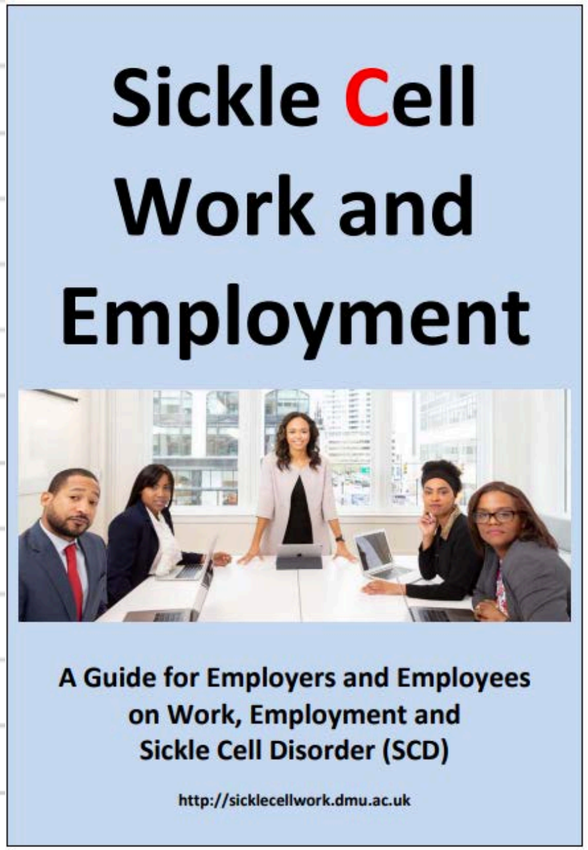
SICKLE CELL DISORDERS

HOW CAN THE WORKPLACE SUPPORT?



Leaflet available online:
https://sicklecellwork.dmu.ac.uk/wp-content/uploads/2019/10/PDF-Version_Guide-To-Sickle-Cell-and-Employment-Version_1.1_2019.pdf

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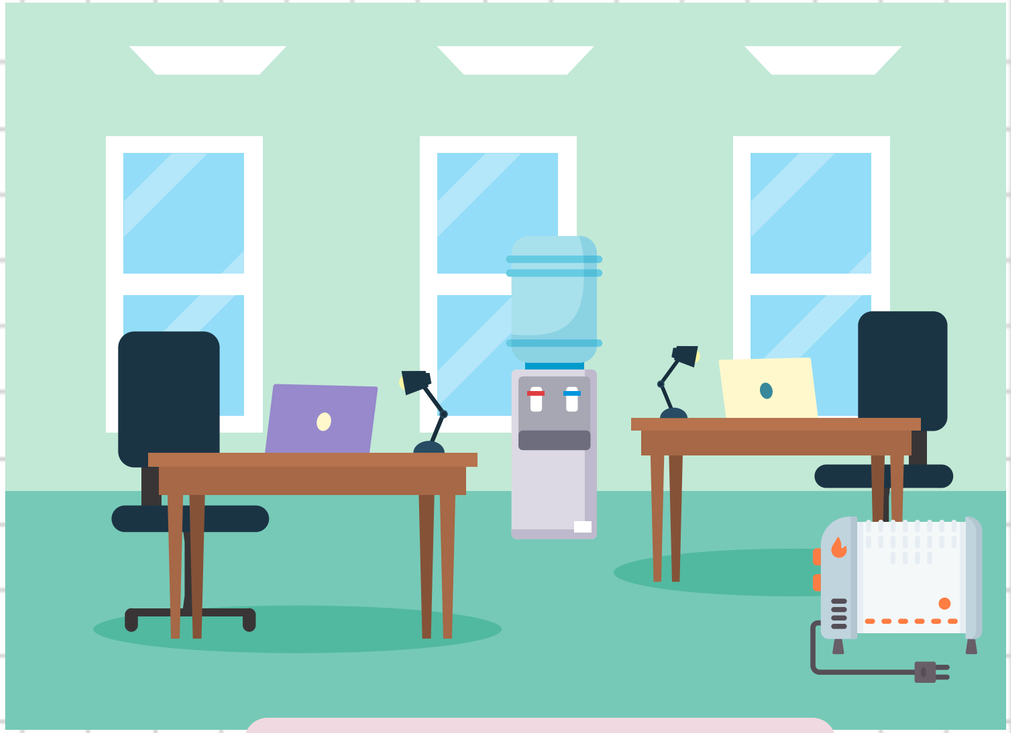
SICKLE CELL DISORDERS

HOW CAN THE WORKPLACE SUPPORT?

Drinking water readily available

No restriction on toilet breaks as need to pass urine more frequently



Desk in a warm part of the office, away from draughty windows

Consider providing a portable heater if office environment is cold

Optimise office conditions

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SICKLE CELL DISORDERS

COMING TO WORK

Flexi-working can help individuals to attend hospital appointments

Offer parking near work/on site to minimise walking distance for those with mobility issues or pain

Consider impact of public transport on infection risk and cold exposure

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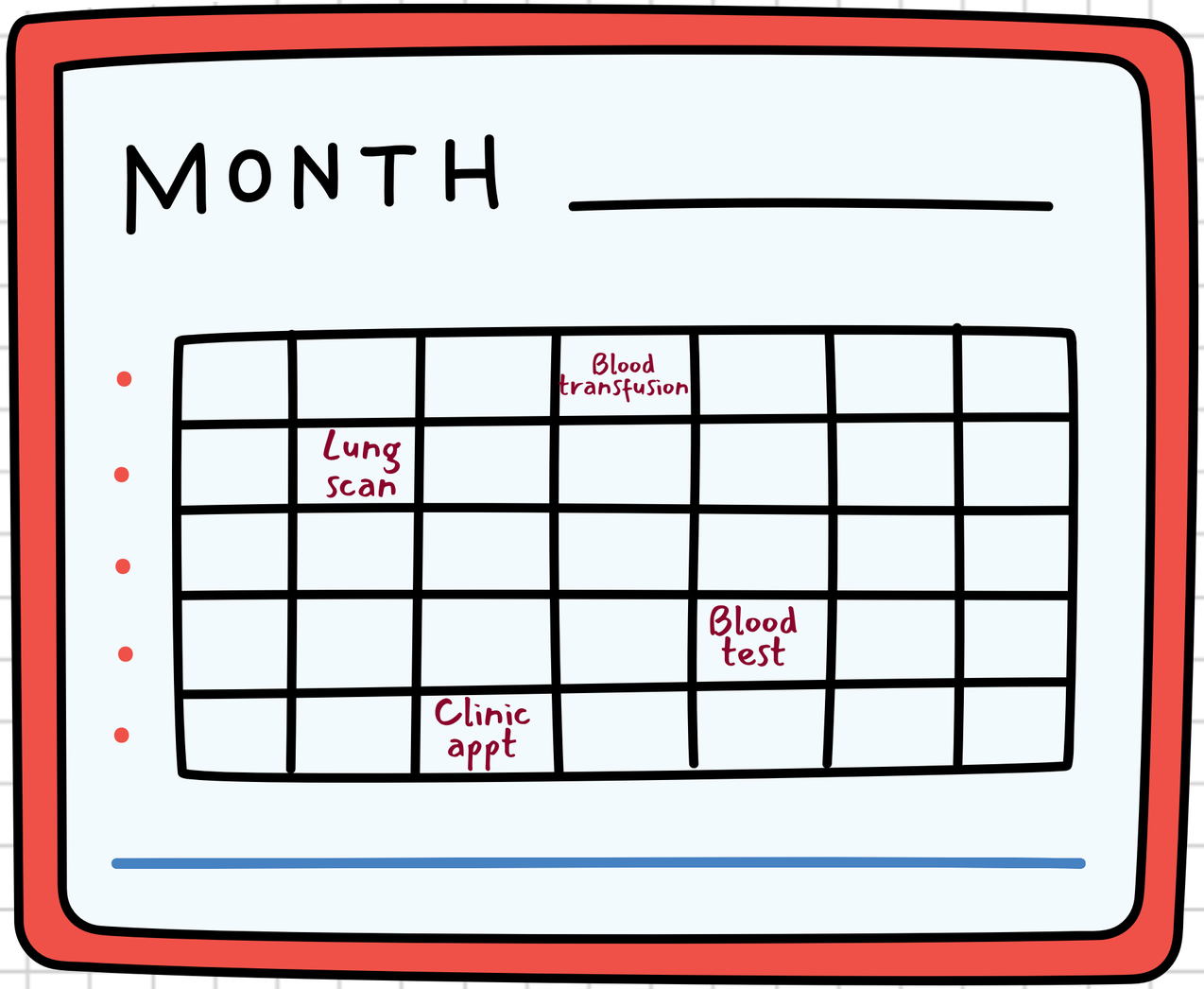
SICKLE CELL DISORDERS

APPOINTMENT BURDEN

Patients often have a high volume of hospital appointments to monitor their health.



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SICKLE CELL DISORDERS

WORK SUPPORT PLANS

This is an example from the "Sickle Cell, Work and Employment" booklet

A Framework for a WORKER'S INDIVIDUAL SUPPORT PLAN for Someone with Sickle Cell Disorder

Name: Date of Birth: Workplace(s): Current work group: **Condition 1:** Sickle Cell Anaemia (HbSS) **Condition 2:** **Condition 3:** [People with SCD may develop other long-term conditions] Date of Plan: Review Date: [Suggest annual review]	 Photograph [taken when the person is well and free from pain]
NEXT OF KIN CONTACTS **Contact Name:** **Relationship:** **Contact number:** **Contact Name:** **Relationship:** **Contact number:**	**CONTACT NUMBERS** **Emergency Contact Name:** **Emergency Contact number:** **Hospital Consultant Name:** **Hospital Consultant Number:** **Specialist Nurse Name:** **Specialist Nurse Number:**
RESPONSIBLE IN WORKPLACE: Name: Building/Department: Contact Number:	**WORKPLACE**

REASONABLE ADJUSTMENTS:

Generic
Key worker to ensure that each workstation is aware of importance of following preventive measures: a warm environment, unrestricted access to water and toilet breaks.

Person-Specific Adjustments

<u>Example 1</u>: Provided height-adjustable chair and foot stool to help with necrosis of the hip and post hip-replacement surgery

<u>Example 2:</u>

<u>Example 3:</u>

MEDICATION

A person with SCD may be prescribed opiate-strength medication as part of overall pain management in hospital. Strong medication such as morphine has side effects and the person may have difficulty sleeping, and may have withdrawal-like symptoms such as sweating, being confused, and feeling dizzy. In some cases they may be prescribed reducing doses for a few days post-discharge to wean off the drugs slowly. Hospital staff and GPs can help by ensuring fit notes cover the week following hospital discharge. HR and managers should respect the need for time to recover, as returning to work too soon without recuperation time can lead to a relapse.

PAIN MANAGEMENT

The aim is to strike a balance between responding appropriately to medical emergencies and maintaining an inclusive work environment where a person with SCD is not frequently off work for all episodes of pain. Many people with SCD experience mild pain at work and have developed the resilience to cope with this and carry on. Sometimes if the pain is moderate this may entail self-medicating with painkillers or using individually tailored pain management strategies such as use of hot water bottles, self-massage, or distraction techniques (e.g. watching TV, listening to music). In some cases a period of rest at work in a quiet safe area may be sufficient to recover and even return to work the same day. In other cases being permitted to go home and rest for the remainder of the day and perhaps the next day may (or flexibility such as working from home the next day) can prevent a full sickle cell crisis developing and much greater time off then being required. In cases of severe pain the person may need to go home or refer themselves to hospital. In cases of excruciating pain an ambulance should be called immediately.

NO PAIN					MOST PAIN
	Self-medication or individual techniques	Self-medication or individual techniques plus "time-out" in quiet safe environment	A couple of day's rest at home in order to prevent a worse crisis	Can go home and manage their own referral to a day care unit	Dial 999 for an ambulance. Know which hospital is their main treatment centre.

ONLY THE EMPLOYEE IS AN EXPERT IN THEIR OWN PAIN AND ONLY THE EMPLOYEE CAN SAY HOW SEVERE THEIR PAIN IS AT ANY GIVEN TIME. ALWAYS LISTEN TO THE WORKER.

OTHER PARTICULAR NEEDS/ISSUES

This section can contain information specific to the person's <u>individual</u> condition (for example, information about mental health, strokes, leg ulcers, eye problems, hip problems, priapism, headaches, seizures, deep vein thrombosis or other possible complications of sickle cell disorder).

Stakeholders in drawing up a Workers Individual Support Plan [WISP]

Name of Person:	Signature:	Date:
Occupational Health:	Signature:	Date:
Sickle Cell Specialist Nurse:	Signature:	Date:
Human Resources Manager:	Signature:	Date:
Union Representative:	Signature:	Date:

Space for a relevant list. If requested by the worker this might include a list of medications they have been prescribed. Alternatively it might be space to note which relevant staff have attended a Sickle Cell Awareness Education Session:

e.g. Name of Prescribed Drug e.g. Name of Staff Member	Dosage Date of Attendance

Space to include examples of good practice developed by the workplace:

Illustrative Example 1

[Name of Employer] has joined the Disability Confident Employers Scheme to embed continuous improvement in disability practice into the organization

Example 2

.....

.....

Example 3

.....

.....

WE HAVE COVERED...

- All about red cells
- The background of sickle cell disorders
- How sickle cells affect the body (complications)
- Helping to prevent sickle-related complications in the workplace

ANY QUESTIONS?
THANK YOU!

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DISCLAIMER: These slides are for education purposes only.
Speak to your doctor if you have any questions relating to your health.