# Sickle Cell Treatment Guide

***Medicines, Modern Therapies and Supplements Explained Clearly***

**DR LEWIS THOMAS, MBBS, MRCGP**

**WHAT EVERY WARRIOR NEEDS TO KNOW**

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## Disclaimer

This book is provided for educational purposes only. It is not intended to replace the advice of your physician, specialist, or any qualified healthcare provider.

The information in this guide reflects both medical training and lived experience with sickle cell disease. Every effort has been made to ensure the content is accurate and evidence-based at the time of publication. However, treatments and recommendations may change as new research and clinical guidelines evolve.

If you have sickle cell disease or are caring for someone who does, always consult your healthcare team before making decisions about medications, therapies, or supplements. Never disregard professional medical advice or delay seeking it because of something you have read in this book.

The author cannot accept responsibility for any loss, injury, or outcome resulting from the use of the information contained herein.

# Contents

# Author's Note

The threat of sickle pain isn't something to be taken lightly. Whether you've suffered it yourself or watched a loved one endure it, the experience leaves a deep mark. It triggers something in the brain that makes it impossible not to think about every possible way to prevent and relieve it.

But with so much conflicting information about treatments and supplements, how do you know what to trust? Perhaps you've had bad experiences with insensitive doctors and felt they didn't have your best interests at heart. On the other hand, maybe you've doubted stories of fellow sickle cell warriors who say they've healed themselves through herbs or prayer alone. Either way, it's natural to ask questions, but when the answers are hard to find or impossible to understand it's incredibly frustrating. At the end of the day, all you really want to know is simple: *What works? And is it safe for me?*

I was only 12 years old when I first began asking those questions. It was the first sickle cell crisis I could remember, and it was a pain more severe than anything I'd ever imagined was possible to live through. I thought death must be close and if it stops the pain I'll go gladly. But as I lay there considering my mortality, I realised just how much I didn't know. I think on a subconscious level that was the start of my journey to understand the metaphysical aspects of the universe. But at the time all I wanted was to know everything I could to avoid ending up in hospital again. I was drawn to study medicine and become a doctor. I needed the rules, structure and evidence that science provided for my questions.

As I've grown older, I've also become more spiritually aware of my purpose in life and where sickle cell fits in. I trained as a doctor for 10 years – from medical school through postgraduate training – and then practiced for 5 years as a fully qualified GP. Having gained knowledge and understanding from both patient and doctor perspectives, I now want to give back. Today, I dedicate my work to empowering sickle cell warriors through education and coaching. Because sickle cell is painful, but knowledge is power – and I've seen the difference it's made in my own life.

So now I want to share that knowledge with you, so you can keep yourself or your loved one healthy enough to fulfil their own purpose. I hope you can be at ease, trusting that I've written this guide in your best interests – because your best interests are also mine.

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## Why I Wrote This Guide

> *"If I'd have known this 10 years ago I might not have had a stroke"*
> - anonymous sickle cell warrior

A couple of months ago I built up the courage to talk about sickle cell on Tiktok. I filmed myself talking about my experience with a sickle cell medication called hydroxyurea. The idea that someone would see it and find it useful motivated me to post.

What followed was a response that completely overwhelmed me. So many people engaged seeking answers and reassurance from me or just wanted to thank me for sharing what I knew. And it turns out I knew a lot, but I'd taken it for granted because I'd never been in contact with this many sickle cell warriors and their advocates at one time. It felt like I'd opened a virtual medical clinic for a moment. Although I'm not legally giving medical advice, I *am* giving knowledge and perspective that allows people to make informed decisions and feel more confident.

I believe the role of a doctor isn't just to write a prescription and call it a day. A prescription alone is useless if the patient never takes it. I learnt this first-hand as a General Practitioner (GP): when people don't understand what a medication does or why it will help them, they're often too afraid to put it in their body. That's why I took pride in communicating with patients in straightforward, relatable terms.

It's kind of like learning to shoot a basketball. Anyone can throw the ball at the hoop, but unless you follow through with that flick of the wrist, it's unlikely to spin properly and land in the net. Prescribing medication without explanation is like

throwing the ball and hoping for the best. Taking the time to counsel and explain is the follow-through - it's what gives the shot the best chance of swishing through the net.

And that's even more important in sickle cell. Too often, people are left piecing together information from rushed clinic appointments, mixed messages on social media, or medical jargon that feels impossible to unpack. Without clarity, it's easy to lose trust and avoid taking something that could genuinely help. The lack of clear, trustworthy information leads to doubt:

*"Am I missing something? Am I making the wrong choice? What if I'm not doing enough to protect myself?"*

That's why I've written this guide, to provide that missing follow-through; the clear, relatable explanations that give you confidence to make decisions and get the most from your treatment.

I want to give you a clear, honest, and practical overview of the treatments and supplements that are most relevant to sickle cell today. I'm not going to overwhelm you with irrelevant scientific detail that serves no purpose other than to sound smart. But I do want to go deep enough so that you can understand everything for yourself and don't have to blindly accept what I say.

There's hardly a surplus of educational sickle cell books out there. So this will at least be much needed additional awareness for the commonest serious genetic condition in the world. At most it'll be an indispensable guide for sickle cell warriors all over the world to arm themselves with knowledge in the fight for health equality.

Think of this Ebook as a **bridge** between medical knowledge and everyday life.

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## Who This Ebook Is For

This guide is for anyone living with sickle cell (their loved ones, carers and support networks) or anyone that cares for someone with the condition. You might be:

- Looking for reassurance about a medication you've been offered.
- Curious about whether a supplement could really help.
- Or simply trying to understand sickle cell treatment better so you feel more in control.

Wherever you are on the journey, this guide is here to support you.

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## What to Expect Inside

➔ **The science made simple:** clear explanations of what sickle cell does to the body, without the heavy jargon.

➔ **Medications that make a difference:** Covering the standard daily tablets, IV drip options and even options to cure sickle cell completely.

➔ **Managing pain step by step:** my "Sickle Cell Pain Ladder," combining medical knowledge with my lived experience.

➔ **Supplements that support your health:** Vitamin D, Omega-3, Zinc, and others that may give you an extra edge.

➔ **Risks explained:** an honest but balanced look at side effects and complications, so you know what's worth worrying about and what's not.

➔ **Personal insights:** sprinkled throughout, where they add context and perspective from my own journey as both a doctor and a sickle cell warrior.

## How to Read This Book

The best way to read this book the first time is from start to finish. Many of the medical concepts build on each other, so by following the chapters in order, you'll get a clearer understanding of how each treatment and supplement fits into the bigger picture.

To avoid repeating myself in every section, I've explained certain key concepts once, then linked back to them where relevant. If you decide to skip ahead to a particular treatment, don't worry – you'll find hyperlinks to any terms or explanations you might need, so you can jump straight to the background information.

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## A Note of Reassurance

This ebook is here to empower you with knowledge, but it's not a replacement for your healthcare team. Every person with sickle cell is unique, and the best medical decisions are always made in partnership with your haematologist or registered physician.

What I can give you is clarity, context, and confidence – so when you sit in that clinic room, you'll feel ready to ask the right questions and understand the answers you're given.

**By the end of this ebook, my hope is that you will:**

- Feel less anxious about your treatment choices.
- Have a clearer picture of the medications and supplements that matter most.
- And feel empowered to take an active role in your own health.

This book is about giving you back a sense of control. Because when you understand your options, you can make better choices for your health, your energy, and your life.

So, let’s get started.

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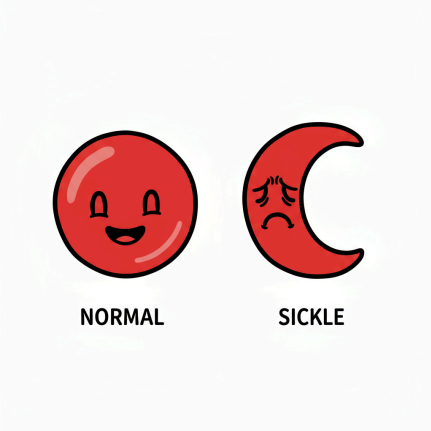
# How Sickle Cell Causes Problems

In order to understand the treatments of sickle cell we need to understand how it causes disease. Below is a brief summary of what you need to know before we dive into the treatments.

## Red blood cells get stuck and block blood flow.

Normal red blood cells have a round, smooth, flexible shape which helps them pass smoothly through blood vessels without issues.

In sickle cell the red blood cells take on a sickle shape which is rigid and inflexible. Because of this they are prone to getting stuck as they pass through tiny blood vessels in the body.



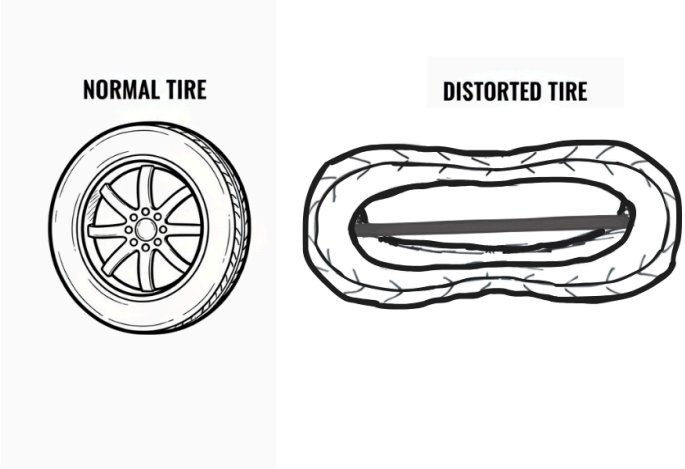

## So why do they change shape?

Red blood cells are made of a red protein called haemoglobin. Haemoglobin is responsible for carrying the oxygen within the red blood cell.

The genetic mutation that causes sickle cell disease results in the production of an abnormal form of haemoglobin, called “sickle haemoglobin.” This creates two major problems:

1. It isn't very efficient at carrying oxygen.
2. When it's not carrying enough oxygen – for example when oxygen levels in the body drop, it sticks together in long chains. These chains distort the normally round red blood cell into a rigid, sickle shape.

Think of it like a car tyre. Normally, a tyre keeps its round shape because the inner spokes are evenly balanced. But imagine if, instead of a set of round spokes, there was a single long metal rod stretching across the tyre. The tyre would bulge and bend into an awkward oval instead of staying round. That's essentially what happens inside red blood cells - the sickle haemoglobin lines up into stiff rods that push the cell out of shape.




## Sickle Cell Crisis

When sickled red blood cells get stuck in a traffic jam inside small blood vessels, oxygen can't reach the body's tissues. This often happens in the bones and joints, which are full of tiny vessels. The lack of oxygen creates an excruciating pain called *ischaemia*.

To put it into perspective: a heart attack is caused by blocked blood vessels stopping oxygen from reaching part of the heart muscle. Now imagine that same type of oxygen-starved pain, but concentrated inside your bones and joints. That's what a sickle cell crisis can feel like.

## To summarise

Red bloods cells change shape, get stuck in small blood vessels, block blood flow and prevent oxygen supply. This is the basic process that causes a range of different problems in sickle cell disease. That's all you need to know at this point. I'll introduce more details, concepts and problems in the context of the medications, therapies and supplements we're about to discuss.

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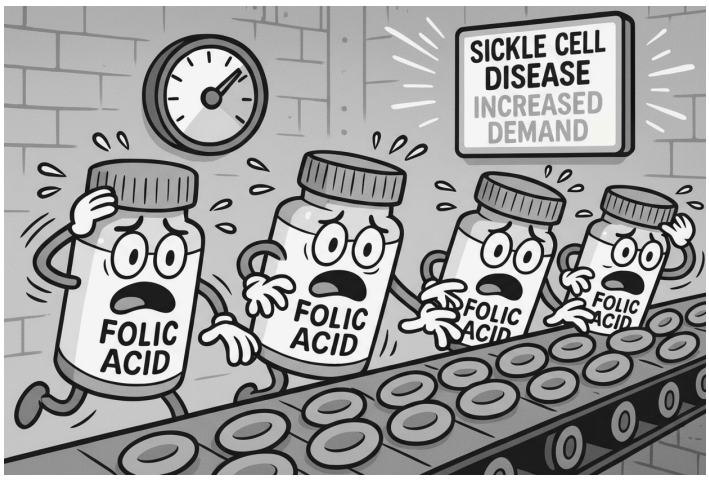
# Section 1: Regular preventative Medication

1) Folic Acid
2) Penicillin
3) Hydroxyurea
4) Endari
5) Why Voxelator (Oxybryta) was banned

## Folic Acid

Folic acid helps your body make new red blood cells. In sickle cell, red blood cells break down much faster than normal. This is because the abnormal sickle haemoglobin damages the cells, so instead of living for 120 days, they only survive about 20 days. This constant breakdown leads to anaemia because your body simply can't make enough new cells to replace all the lost ones.

This is where folic acid becomes important. Folic acid is a key ingredient your body uses to build DNA in new cells. The production of red blood cells takes place in the bone marrow. Imagine this as a red blood cell making factory, where folic acid is the fuel that keeps the machines running. Without enough fuel, the factory slows down and you may become more anaemic. That's why people with sickle cell need higher levels of folic acid to keep the factory working at maximum speed.




You can get folic acid from foods like leafy green vegetables, beans, and fortified cereals. But because the demand for new red blood cells is so high in sickle cell, diet alone usually isn't enough.

That's why it's recommended to take a **5 mg folic acid tablet every day**.

This helps ensure your bone marrow never runs out of the raw material it needs to keep producing healthy red cells. To put this in context, the standard recommended daily amount of folic acid for the general population is just **0.2–0.4 mg (200–400 micrograms)**, that's more than **10 times lower** than what sickle cell warriors need.

Folic acid is very safe, with few side effects. The only caution is that very high doses can mask a vitamin B12 deficiency, which is rare but worth being aware of. For most people, though, daily folic acid is one of the simplest and most effective things you can do to support your blood health.

**Parents' Note**

**Folic Acid**

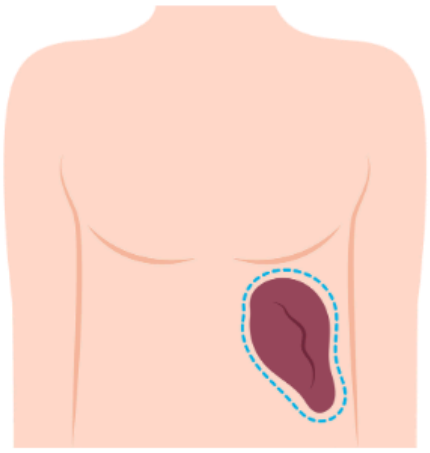
- **When it's started**: From diagnosis in childhood.
- **Typical dose for children**: Same as adults, **5 mg once daily**.
- **How long it's used**: Ongoing - lifelong in most cases.
- **Safety**: Very safe; higher dose than the general population is expected and necessary.
- **Extra tip**: Folate-rich foods are healthy, but supplements are essential to meet demand.

## Penicillin

Penicillin is an antibiotic that protects the body from harmful bacteria. In sickle cell, it's taken regularly to prevent severe bacterial infections.

In order to explain why this is required in sickle cell, let me introduce the spleen.

**Anatomical location of Spleen**



**Spleen is very vulnerable to sickle cells**

Your spleen sits in the upper left of your belly. It plays a key role in your immune system by helping to filter bacteria out of your blood and by making white blood cells to fight infection.

But in sickle cell, the spleen often doesn't work properly. The tiny blood vessels inside it are prone to getting blocked by sickled red cells, which can damage the spleen over time. This means your body loses one of its best natural defences against infection. Penicillin is prescribed as an extra layer of protection, particularly against dangerous bacteria like *pneumococcus*. This is the germ that can cause serious illnesses such as pneumonia, meningitis, and sepsis (blood poisoning). These infections can come on suddenly and be life-threatening in people with sickle cell, especially young children, which is why daily penicillin is so important.

## Are there any side effects?

Most people tolerate penicillin well. But like any antibiotic, it doesn't always agree with everyone. This can look like uncomfortable but relatively harmless stomach issues - nausea, vomiting, diarrhoea, or constipation. These are what we call intolerances.

A small number of people are allergic to penicillin, with symptoms like rashes, itching, or swelling. Anaphylaxis is a rare but severe allergic reaction that you would notice immediately. Wheezing, difficulty breathing, throat swelling, or sudden lightheadedness and fainting are all red flags that require emergency treatment.

If someone with sickle cell is allergic to penicillin, doctors usually prescribe another antibiotic called **erythromycin** instead. This works in a similar way to protect against bacteria such as *pneumococcus*, so the person is still protected from serious infections.

## How long do you need it for?

Children with sickle cell are usually started on penicillin soon after birth and continue until at least age 5. This is because young children are at the highest risk of severe infections. After age 5, doctors may decide to stop penicillin if the child is healthy and has built up more protection – both from routine vaccinations and from the natural development of their immune system.

The decision to continue beyond age 5, or to stop as an adult who is already taking it, should always be made in consultation with your doctor. They will help you weigh up the benefits of ongoing protection against infection with the downsides of long-term antibiotic use, such as resistance or the inconvenience of daily tablets.

**Parents' Note - Penicillin**

- **When it's started**: From birth.
- **Typical dose for children**: Under 5 years it's based on age/weight. Over 5 years is same as adult dose 250mg twice daily.
- **How long it's used**: By 3 months until at least age 5; longer in some cases, decided by doctor.
- **Safety**: Very safe, well-tolerated.
- **Alternatives**: Erythromycin if allergic.
- **Extra tip**: Daily penicillin is one of the simplest and most effective ways to protect young children with sickle cell from life-threatening infections.

# Hydroxyurea

**My experience with Hydroxyurea**

I've taken hydroxyurea for the past 10 years. While on it, I was able to live a relatively normal life without severe crises or hospital admissions. Last December 2024, I decided to stop taking it to see if I could manage without. Within 3 months, I had my first admission in 10 years. That was the moment I realised I'd rather take a tablet every night and have a blood test every 3 months than end up back in a hospital bed.

Hydroxyurea also known as Hydroxycarbamide stops red blood cells from sickling. It's one of the longest established treatments for sickle cell disease and helps to:

- Prevent sickle cell crisis
- Reduce the severity of pain crisis
- Reduce risk of strokes
- Reduce risk of Acute Chest Syndrome
- Reduce build-up of damage to your organs from sickle cell over time

## Hydroxyurea works by increasing Fetal Haemoglobin

To fully understand how hydroxyurea is so beneficial in sickle cell disease we need to talk about Fetal Haemoglobin.

## What is Fetal haemoglobin?

Fetal haemoglobin (HbF) is a special type of haemoglobin made by babies in the womb. It's excellent at carrying oxygen because its job is to pull oxygen from the mother's blood supply. Once babies are born and can breathe on their own, HbF is

no longer needed, so the body gradually switches it off around 6 months of age. From then on, adults mostly produce haemoglobin A - or in sickle cell, haemoglobin S.

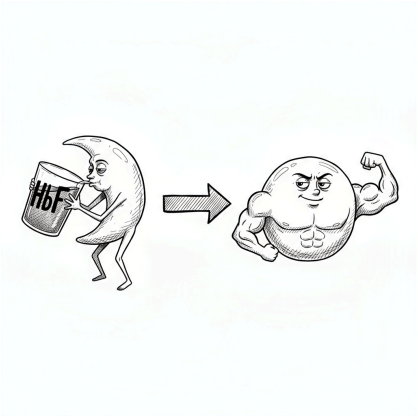
If we ranked haemoglobin types in terms of oxygen-carrying ability, it would go like this:

1. Fetal Haemoglobin (HbF) - strongest
2. Adult Haemoglobin (HbA)
3. Sickle Haemoglobin (HbS) - weakest

In sickle cell, symptoms often start once HbF has disappeared and HbS takes over.

## How does Fetal Haemoglobin improve sickle cell?

HbF is highly resistant to sickling. When red blood cells contain enough HbF, they stay round and flexible instead of turning into the sickle shape. This means HbF can ‘compensate’ for HbS - protecting the red cells and reducing sickle cell symptoms.




## How does Hydroxyurea increase Fetal Haemoglobin?

In sickle cell hydroxyurea acts on the bone marrow where all blood cells are made. It has the effect of slowing down the production of all blood cells including white blood cells and platelets which we’ll discuss in a moment.

Remember in sickle cell the body is already trying to produce as many red blood cells as possible to keep up with the ones being destroyed. So then the question becomes how does slowing this down help? Well the reason for this is to force the body into playing its trump card. That's right; the body still has the ability to produce fetal haemoglobin and we are tricking it into doing so.

Think of it like this. Your body is a company that's being put under pressure to produce results. This triggers an all hands on deck experience where the body calls back its most effective staff (Fetal Haemoglobin) from retirement to help out. So a small amount of fetal haemoglobin returns to get more red blood cells made.

## Other benefits of Hydroxyurea

Hydroxyurea reduces the number of white blood cells made and this can help to reduce the amount of inflammation in the blood, which makes it less sticky; thereby reducing the chances of sickled red blood cells getting stuck.

## How to start taking Hydroxyurea

Usually Hydroxyurea is only available via prescription from your Haematology specialist doctor. However in some countries Hydroxyurea is available to buy without a doctor's prescription.

**You should avoid starting hydroxyurea without the supervision of a haematologist.**

Before starting hydroxyurea, your doctor will take some baseline blood tests to check your red cells, white cells, platelets, and kidney and liver function. Once you begin, your blood will usually be checked every 2-4 weeks while the dose is slowly increased to find the safest and most effective level for you.

After your dose is stable, blood tests are usually needed every 2-3 months to make sure everything stays safe. This monitoring is one of the reasons hydroxyurea is considered a reliable and trusted treatment; because your healthcare team is checking regularly that your body is responding well and staying healthy.

You'll only start to feel the benefits of hydroxyurea after taking it **consistently for 3 months**. This is how long it takes for the hydroxyurea to influence the bone marrow into making enough Fetal Haemoglobin containing red blood cells. If you forget or decide to take breaks from it, it will not be effective.

## Are there any side effects?

Yes, like all medications, hydroxyurea can have side effects.

To help give you the best idea of what to be aware of and look out for I've organised the side effects into the 2x2 table below. It's important when considering side effects to understand Common vs Uncommon and Harmless vs Serious.

| | Harmless / Mild | Serious (need medical attention) |
|---|---|---|
| **Common** | -Darkened nails or skin<br>-Mild nausea or stomach upset<br>-Temporary tiredness<br>-Mild drops in blood counts (usually no symptoms, it's picked up on routine tests) | More significant drop in blood counts → may cause unusual tiredness, easy bruising, or more frequent infections (fever, sore throat) |
| **Uncommon** | - Mouth ulcers<br>- Mild hair thinning<br>- Mild skin rash | -Allergic reaction (rash, swelling, breathing difficulty)<br>- Severe infection due to very low white cells (signs: fever >38°C (100.4°F) , chills, shortness of breath)<br>- Liver changes (rare, usually no symptoms but picked up on tests)<br>- Worsening of leg ulcers (in people who already have them, or new ulcers developing in some cases) |

Most people tolerate hydroxyurea well. The majority of side effects are **mild and harmless**. The **serious ones are very uncommon**, and your healthcare team monitors for them with **regular blood tests**. This means problems can usually be spotted and managed early, long before they become dangerous.

## What about fertility?

Hydroxyurea does **not** cause permanent infertility.

In men, it can **temporarily lower sperm count or quality** while they are taking it. Research shows this usually improves within **3-4 months of stopping the medication**. In fact, several studies have reported men successfully fathering healthy children even while on hydroxyurea. It's also worth noting that in some cases, **severe sickle cell disease itself can impair fertility**, even without hydroxyurea - often due to testicular damage from sickling episodes or the effects of chronic illness and iron overload.

In women, there's **no evidence that hydroxyurea causes permanent infertility**. Most women are able to conceive after stopping it. However, severe sickle cell disease itself can sometimes affect fertility in women too - for example, by lowering ovarian reserve or increasing complications during pregnancy.

Because research is still ongoing, doctors usually recommend **sperm banking for men** or discussing **fertility preservation options for women** before starting hydroxyurea, where possible. This isn't because it's known to cause permanent infertility, but more as an extra precaution to protect future family planning options. There's no need to worry if you'd already started hydroxyurea before being able to preserve fertility.

## History of Hydroxyurea and Cancer myth

I want to address Hydroxyurea's history and the rumours surrounding its origin as a chemotherapy drug. Now that you have the context behind how hydroxyurea improves sickle cell I hope that you'll be reassured by the following summary.

**1950s** - Hydroxyurea was discovered. Scientists found it could slow down DNA replication in lab experiments. This led to public fears that it might 'damage DNA' or cause mutations.

**1967** - Hydroxyurea was approved as a chemotherapy drug for certain types of leukaemia. In this setting, the goal was to slow down the spread of cancer cells. This was nothing to do with causing cancer. But the 'chemo' label created lasting fear.

**1980s** - Doctors noticed that cancer patients with sickle cell who were on hydroxyurea had far fewer pain crises and milder symptoms. Research confirmed this was due to a rise in Fetal haemoglobin.

**1990s** - Large trials began in people with sickle cell at much lower doses than those used in cancer. The landmark **1995 MSH Study** showed hydroxyurea reduced pain crises, acute chest syndrome, and the need for transfusions.[1]

**Today** - More than 30 years of follow-up studies show **no increased risk of cancer** in sickle cell warriors taking hydroxyurea.

Hydroxyurea's label as a 'chemo drug' is old news. In sickle cell it's given at much lower doses, with a completely different goal. I understand how the idea of scientists trying out a medication on black people can raise suspicions. Because black people have been abused and mistreated by racist medical institutions in the past. But trust me, hydroxyurea is not another trick or conspiracy against black people. After 30 years of long-term safety there's no further need for suspicion. This is a vital medication for sickle cell that everyone should try if able to access it safely under medical supervision.

**Parents' Note - Hydroxyurea**

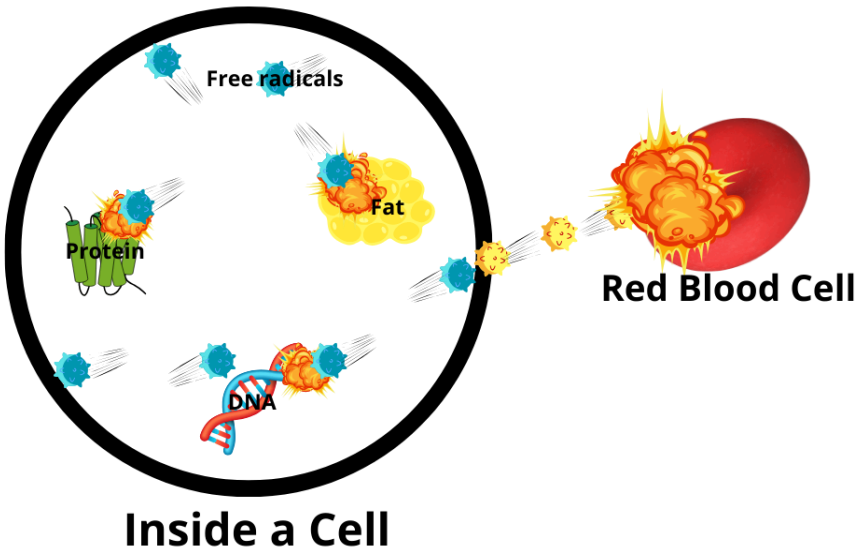
- **When it's started:** Hydroxyurea can be prescribed for children as young as 9 months old if they are having frequent sickle cell problems.
- **Typical dose for children:** The dose is based on the child's weight and adjusted gradually. It is not the same for everyone, and doctors monitor closely with regular blood tests.
- **How long it's used:** Hydroxyurea is usually a long-term treatment. Children may need to continue it throughout life, as long as it is working well and side effects are minimal.
- **Safety:** Hydroxyurea has been studied in children for over 20 years. It is safe when monitored, and most children tolerate it well.
- **Fertility:** There is no evidence of permanent infertility in children treated with hydroxyurea. As an extra precaution, teenage boys may be offered sperm banking before starting.
- **Extra tip:** Parents sometimes worry about the "chemotherapy" label. In sickle cell, hydroxyurea is given at a much lower dose than in cancer, and decades of research show it is safe and effective for children.

# Endari

Endari boosts **anti-oxidant** production by sickle cells.

## Oxygen free-radicals and Anti-oxidants

When our body's cells use oxygen to make energy, not all of it gets used up cleanly. Some oxygen escapes in a damaged, unstable form called ***Free Radicals***. Imagine them as wild pinballs bouncing around inside cells, crashing into proteins, fats, and even DNA.




In sickle cell red blood cells are vulnerable to damage from free radicals. When they meet it can result in sickling and destruction of red blood cells. Free radicals get to red blood cells in 3 different ways

1. Via damaged body cells adding destructive by-products to the blood stream.
2. When white blood cells release free radicals to fight germs, some of them hit red blood cells as innocent bystanders. In sickle cell, white cells are often overactive and release free radicals even without an infection, creating extra damage.

3. Sickled haemoglobin can leak iron, and iron drives chemical reactions that generate even more free radicals inside the cell.

With all this free radical craziness known as '**oxidative stress**' red cells become weaker, more likely to sickle and more likely to break apart.

## How Endari helps

The body fights free radicals using antioxidants. Antioxidants neutralise free radicals safely, like putting out sparks before they cause fire. The body makes antioxidants naturally, such as its most powerful one, **glutathione**. It also gets anti-oxidants from foods like fruits and vegetables (vitamins C and E).

But in sickle cell, the amount of oxidative stress is so high that natural antioxidant defences often get overwhelmed.

**Endari (L-glutamine)** helps by boosting the body's production of glutathione. With more antioxidant protection, red blood cells:

- Survive longer (reducing anaemia),
- Are less sticky and more flexible (so they pass through blood vessels more easily)
- Are less likely to sickle.

## How is it taken?

It's taken as an oral powder sachet dissolved in water.

Endari is currently licensed in the USA and some parts of Africa. It is not yet available in Europe, which seems to be more due to regulatory processes and research approvals than any problem with its effectiveness.

Studies and patient reports show that Endari can work well, particularly for people who cannot take hydroxyurea or do not tolerate it.

## Can I just buy L-glutamine supplements instead of Endari?

L-glutamine is the same ingredient found in Endari, and you'll often see it sold online as a high-strength workout powder. The difference is that **Endari is a prescription medicine**:

- It's manufactured to strict medical standards.
- It's been tested in clinical trials for sickle cell disease.
- The doses used (10-30 g per day depending on weight) are carefully measured.

Over-the-counter powders aren't tested in sickle cell patients, may vary in purity, and aren't guaranteed to be safe at high doses. That's why doctors recommend prescription Endari - because we can be certain it works and is safe at the right dose. However we will explore L-glutamine supplements more in the supplements section.

## Recommended dose (by weight):

- Children 5-10 years: 5-10 grams twice daily
- Older children and adults: 10-15 g twice daily (up to 30 g/day)
  The powder is mixed with food or drink.

## Are there any side effects?

Most people tolerate Endari well. Possible side effects include:

- Constipation
- Nausea
- Headache
- Cough

- Abdominal pain
- Pain in back, chest, or arms/legs

These are usually mild. If they persist or become troublesome, your doctor can review your treatment.

**Parents' Note - Endari**

- **When it's started:** Endari is approved for children **aged 5 years and older** with sickle cell disease.
- **Typical dose for children:** Weight-based.
  - 5–10 g twice daily for younger children (5–10 years).
  - 10–15 g twice daily for older children and adolescents (up to 30 g/day).
- **How it's taken:** A powder mixed with food or drink, usually twice a day.
- **Safety:** Generally well tolerated. Side effects are usually mild, such as constipation, nausea, headache, cough, or stomach aches.
- **Alternatives:** Over-the-counter L-glutamine powders exist, but these are not tested in sickle cell and don't meet medical quality standards. Prescription Endari is preferred because it's proven safe and effective.
- **Extra tip:** Endari can be a helpful option for children who don't tolerate hydroxyurea well, but always discuss with your child's haematology team before making changes.

## Why Voxelator (Oxybryta) was banned

Voxelator was an oral tablet medication that increased haemoglobin levels by reducing sickling.

Voxelotor was helping a lot of people with sickle cell disease, but in some patients it was linked to serious complications and deaths. Because of this, regulators and Pfizer decided it was too risky to keep supplying it, and it was withdrawn worldwide in 2024.

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# Section 2: Intravenous therapies

1) Blood transfusions

- Simple blood transfusions
- Exchange transfusions
- Risks and side effects

2) Iron Overload explained

3) Crizanilizumab (Adakveo)

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## Simple blood transfusions

A blood transfusion gives you a top up of healthy red blood cells to increase your haemoglobin level when it drops too low. Haemoglobin is vital because it carries oxygen around your body to keep your organs and tissues alive and working properly.

Simple transfusions are most often used in **emergency situations**, for example:

- Heavy blood loss,
- Severe anaemia,
- Complications in pregnancy.

Sometimes they're also used **routinely** - for instance, if you're due to have surgery where blood loss is expected.

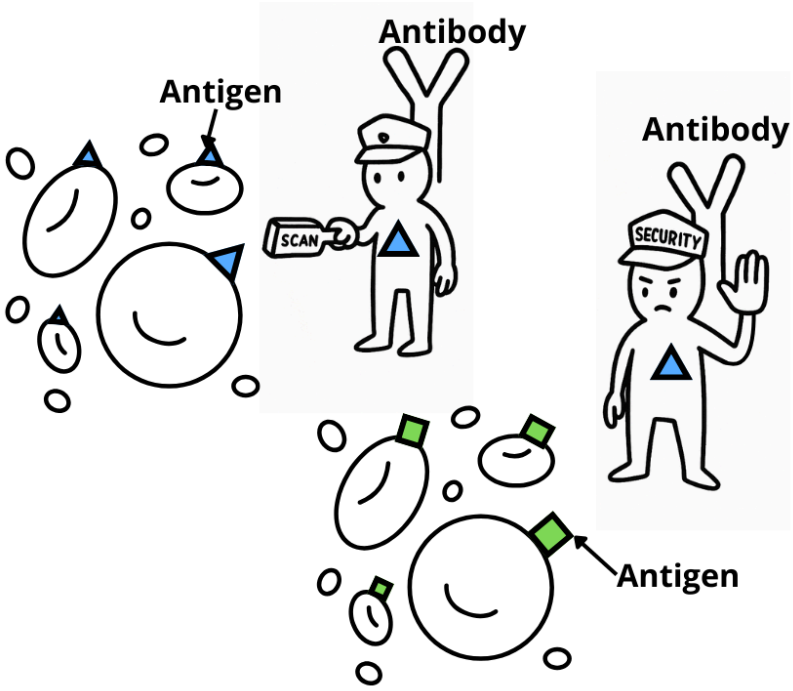
### How does it work?

Before a transfusion, your blood is tested to confirm your **blood group**. This is vital because the blood you receive must be matched to yours - otherwise your immune system may attack it.

Your blood group is determined by **antigens**. These are proteins that sit on the surface of your red blood cells. Think of them as ***ID tags*** that tell your immune system, *"**I belong here.**"*

Your immune system produces **antibodies**, which are like security guards. If they spot red cells carrying antigens that don't match your own, they treat them as invaders and attack. This is what happens in a **transfusion reaction**.

To prevent this, donated blood is carefully matched to your blood group so that your antibodies won't see it as foreign.




## ABO and Rhesus Blood Groups

I've included this overview of blood groups as extra detail for anyone who's interested in the antigens and anti-bodies that determine each blood group. But it's not really necessary to know this as your blood group is screened automatically each time you have a blood transfusion.

| Blood Group | Antigens on Red Blood Cells | Antibodies in Blood | Rh factor |
|---|---|---|---|
| **A+** | A antigen + Rh(D) antigen | Anti-B | Rh positive |
| **A-** | A antigen | Anti-B, Anti-D | Rh negative |
| **B+** | B antigen + Rh(D) antigen | Anti-A | Rh positive |
| **B-** | B antigen | Anti-A, Anti-D | Rh negative |
| **AB+** | A antigen + B antigen + Rh(D) | None | Rh positive |
| **AB-** | A antigen + B antigen | Anti-D | Rh negative |
| **O+** | None + Rh(D) antigen | Anti-A, Anti-B | Rh positive |
| **O-** | None | Anti-A, Anti-B, Anti-D | Rh negative |

## Why transfusions are more complicated in sickle cell

For most people, matching ABO and Rh (positive/negative) blood groups is enough. But in sickle cell, things are more complicated.

That's because:

- There are many other, rarer antigens on red blood cells.
- People with sickle cell who need regular transfusions are more likely to eventually receive blood with one of these rarer antigens, which can trigger antibody production.
- If they receive blood with that rare antigen again, their immune system will attack it, causing a delayed transfusion reaction.

In the past, blood wasn't always screened closely enough for these rarer antigens, and reactions were more common.

Today, things are much safer:

- If you have sickle cell, your blood is usually given **extended matching** - not just ABO, but also other key antigens.
- Labs now screen much more carefully for these rare antigens to reduce the risk.
- Transfusions are monitored closely before, during, and after, to catch any reaction quickly.

So, while transfusions carry risks, they are also **life-saving treatments**. With modern screening and monitoring, they are much safer than in the past.

## Why Black Blood Donation Matters

- Rare antigens are most often shared within the **same ethnic groups**.
- Historically there have been too few Black blood donors and many sickle cell patients received blood from other ethnic groups.

- Over time they've developed antibodies against the rare antigens unique to those groups so are no longer able to receive them.
- With only a limited supply of black donors there's not enough blood for a completely safe match.

Therefore, we need more black blood donors. Every donation directly increases the chances of finding a safe, life-saving match for someone living with sickle cell.

If you're eligible, donating blood could make you the exact match a sickle cell patient has been waiting for.

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## Exchange Transfusions

So far, we've looked at ***simple transfusions***, where healthy donor red cells are added on top of your own. This is useful in many situations, but it also comes with some limits. A simple transfusion raises your haemoglobin, but it doesn't actually reduce the number of sickled red cells already circulating in your body.

That's where **exchange transfusion** comes in. Instead of just topping up your blood, this procedure removes some of your sickled red cells while replacing them with healthy donor red cells at the same time.

Exchange transfusion is usually used for more serious or high-risk situations, such as:

- Preventing or treating strokes
- Severe acute chest syndrome
- Reducing complications during major surgery
- Prevention of serious complications during pregnancy
- When frequent simple transfusions are causing too much iron build-up.

It's essentially a more powerful version of transfusion therapy, designed to not only boost oxygen delivery but also quickly reduce the proportion of sickled red cells in your body.

### How does it work?

There are two ways of doing an exchange transfusion.

1) Automated exchange transfusion

The modern method uses a machine (apheresis) to take out sickled blood and replace it with donor blood continuously. This is quicker, more precise, and reduces iron overload, so it's the preferred option in most specialist centres.

2) Manual exchange transfusion

Blood is removed and replaced in small steps over time. It's less common today, but it may still be used in some hospitals or in countries where the automated machines aren't available. It can also be done in emergencies if apheresis isn't immediately accessible.

## Regular exchange transfusions

If sickle cell is severe, exchange transfusions may be given regularly to prevent complications such as severe pain or stroke. This usually occurs every 4-6 weeks. This timing balances how long donor red cells survive with the need to keep sickled cells at safe levels.

Warriors on a regular exchange transfusion programme often notice a pattern in their energy levels. The week after the transfusion can feel tiring because your body is adjusting to the procedure and the changes in blood volume. Then, towards the end of the cycle, fatigue can creep back in as the donated cells are gradually used up and your own sickled cells take over again. In between, many people describe feeling at their best. This pattern is normal and doesn't mean the treatment isn't working.

## Why do transfusions wear off so quickly in sickle cell?

In healthy people, red blood cells live for about 120 days. But in sickle cell, donor red cells often don't last that long.

That's because the sickle cell environment is tougher on red cells:

- **Inflammation and stress** in the bloodstream make donor cells break down faster
- **Antibodies** from past transfusions can tag donor cells for earlier removal
- **Oxidative stress** weakens cell membranes

- **Storage lesion**: the storage process itself makes donor cells less flexible and quicker to clear

In short, sickle cell creates a harsher environment for red blood cells, so even healthy donor cells don't survive as long as they would in someone without the condition. This is why transfusions need to be repeated regularly to keep up their benefit.

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## Risks and Side Effects of Blood Transfusions

Blood transfusions are life-saving treatments, but like all medical procedures they carry some risks. Most transfusions go smoothly, especially with the extra checks now used for people with sickle cell. Still, it's important to understand the possible side effects and how doctors monitor for them.

### Transfusion Reactions

A transfusion reaction happens when your immune system recognises donor red blood cells as 'not mine' and starts to attack them. This is caused by differences in **antigens** - the 'ID tags' on the surface of red cells.

Reactions can be:

- **Mild**
    - Fever, chills, rash, or itching.
    - These can usually be treated quickly and don't cause lasting harm.
- **Severe (rare)**
    - Shortness of breath, chest pain, low blood pressure, or back pain during or shortly after the transfusion.
    - These require immediate medical treatment.

Doctors reduce this risk by:

- Matching not only ABO and Rh groups, but also many other rare antigens (“extended matching”).
- Closely monitoring patients during and after transfusion.

## Alloimmunisation (antibody build-up)

This is more common in people with sickle cell who need many transfusions. Each time you receive blood, there’s a chance your immune system may form new antibodies against a rare antigen.

- At first, you may not notice anything.
- But in future transfusions, if blood with that antigen is given again, your immune system will attack it - causing a **delayed transfusion reaction**.
- This can make it harder to find safe donor blood in the future.

Extended matching has made this problem less common, but it can still happen.

## Infection Risk

In the past, infection from blood transfusion was a real worry. Today, blood is screened extremely carefully for viruses such as HIV and hepatitis. The risk is now **very, very small** - much lower than most everyday risks.

## Iron overload

Long-term build-up of iron from repeated transfusions (see next section).

Overall the important thing to know is that serious side effects are rare. For most people with sickle cell, the benefits of transfusion far outweigh the risks. Modern

matching, careful monitoring, and regular checks make transfusions much safer today than ever before.

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## Iron Overload

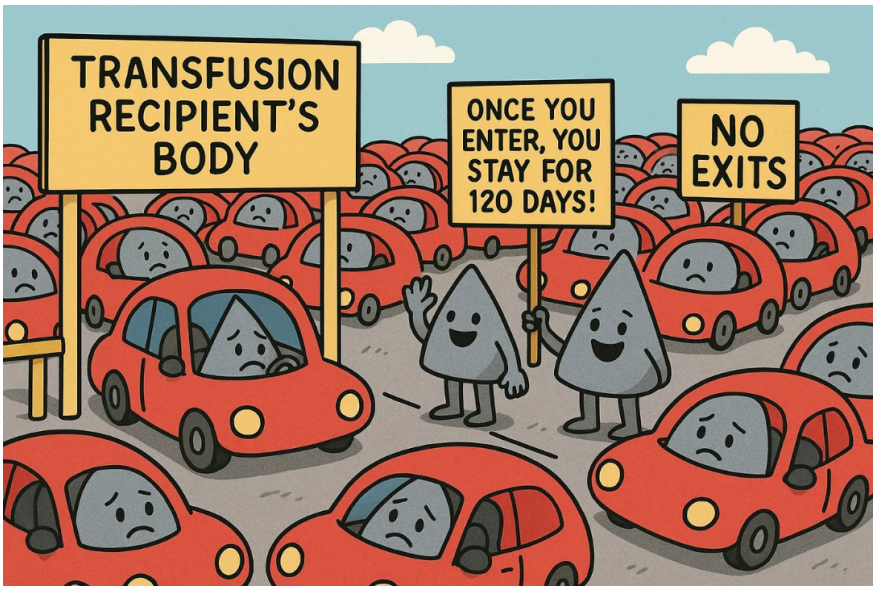
Iron is present in the haemoglobin of red blood cells in every unit of donor blood received. This makes iron overload one of the main long-term risks of having frequent blood transfusions.

Iron is essential for life, we need it to make haemoglobin and carry oxygen, but the body has no natural way of getting rid of **excess iron**. Unlike water or salt, which the body can balance and flush out, iron has no way of leaving.

That means when you keep receiving transfused blood, iron slowly builds up in the body with no way to leave.

Think of your body as a giant car park for red blood cells. Each donor red cell car arrives carrying iron as a passenger. After a while, the cars break down and disappear (the red cells are destroyed), but the iron passengers are still there stranded inside the car park. With every new transfusion, more and more iron passengers are left behind, until the car park becomes overcrowded with iron.

This is how iron overload develops.



## Why does iron overload matter?

When too much iron builds up in the blood, it doesn't just float around, it gets deposited in vital organs, where it starts to interfere with how they work:

- **Heart** → heart failure or abnormal rhythms
- **Liver** → cirrhosis (scarring of the liver)
- **Pancreas** → diabetes

Iron can also affect hormones, joints, and general energy levels. Left untreated, iron overload can cause serious long-term complications.

Review the Deep Dive below for more details.

**Deep Dive: How Iron Overload Damages the Body**

When iron settles into organs it ends up reacting with oxygen and creating those annoying "**free radicals.**" Over time, these free radicals damage cells and cause scarring. Here's what that means in different parts of the body:

**Heart**: Iron in the heart muscle can weaken it (**heart failure**) or upset the electrical system (**irregular rhythms**). This usually only happens after many years without treatment.

**Liver**: The liver is the main iron store. Too much iron here can lead to inflammation and scarring (**cirrhosis**). But regular MRI scans can spot this long before it becomes serious.

**Pancreas**: Iron interferes with insulin production, leading to **diabetes**. Blood sugar checks often pick this up early.

**Hormones:** Iron can affect glands that control growth and hormones, especially the **pituitary gland** (puberty, fertility) and, less commonly, the **adrenal glands** (stress hormones). Children on transfusion programmes are monitored for growth, puberty, and hormone balance.

**Joints & energy**: Iron can stiffen joints and increase oxidative stress, which adds to **fatigue and joint pain**.

## How do doctors check for iron overload?

Monitoring is key, and doctors use two main tools:

- **Blood tests (serum ferritin):** gives a rough idea of body iron levels.
- **MRI scans of the liver and heart:** the gold standard to see exactly how much iron has built up in organs.

Regular monitoring means iron overload can be spotted and treated early, before major problems develop.

## How is it treated? - Iron Chelation Therapy

The only way to remove excess iron from the body is with **iron chelation therapy**. These medicines act like magnets, they grab hold of extra iron and carry it out of the body through urine or stool.

There are three main chelation medicines:

- **Deferasirox (Exjade, Jadenu):** tablets, taken once a day.
- **Deferiprone (Ferriprox):** tablets or liquid, taken multiple times a day.
- **Desferrioxamine (Desferal):** Given as a slow infusion under the skin, often overnight using a small pump**.**

Doctors choose the best one based on your age, health, and personal circumstances.

**Side effects:**

Some people have none. However if they do occur they can include:

- Stomach upset, diarrhoea, or nausea are fairly common.
- Rarely, chelators can affect the kidneys, liver, hearing, or eyesight - which is why regular monitoring tests are essential.

## How long does it take to develop iron overload?

Iron overload doesn't happen after just one or two transfusions. It usually builds up after **dozens of transfusions over several years**. The exact timing depends on:

- How often you're transfused,
- How much blood you receive each time,
- Your body size and storage capacity.

This is why iron levels are checked regularly if you're on a transfusion programme.

## What about exchange transfusions?

You might be wondering about exchange transfusions, since they remove some of your blood at the same time as giving donor blood. Doesn't that mean the risk of iron overload is gone?

Not completely. Exchange transfusions do reduce the risk compared to simple transfusions, because some of your own iron-containing red cells are removed during the process. But the amount of donor blood given is usually **greater than the amount removed**, so you still end up with extra iron in your body.

The good news is that iron builds up much **more slowly** with exchange transfusions than with simple transfusions. That means the risk is lower, but not eliminated. People on regular exchange programmes still need monitoring for iron overload over time.

## Does diet help?

Diet changes can ***support*** treatment, but they cannot remove iron from the body. Only chelation therapy can do that.

**Foods that can *reduce iron absorption***

These are useful to include **with meals** if you have high iron levels:

**Calcium-rich foods** - milk, yoghurt, cheese
→ Calcium competes with iron for absorption.

**Phytate-rich foods** - oats, whole grains, legumes
→ Phytates bind iron and reduce absorption.

**Polyphenol-rich foods** - tea, coffee, cocoa, some herbal teas
→ Especially **black tea** has been shown to cut iron absorption by up to 60%.[2]

**Eggs** - contain **phosvitin**, a compound that binds iron.

**Foods high in oxalates** – spinach, beetroot, nuts, rhubarb
→ Oxalates reduce iron uptake, but note that spinach also contains iron - though it's poorly absorbed.

**Foods and drinks that *increase* iron absorption**

If you're managing iron overload, try to **reduce** these:

**Vitamin C rich foods** - citrus, strawberries, peppers, tomatoes
→ Vitamin C boosts non-haem iron absorption by up to 3-4 times.[3]

**Red meat, liver, poultry, and seafood**
→ These contain **haem iron**, which is absorbed much more efficiently than plant-based iron.

**Alcohol** - especially wine and spirits
→ Increases iron absorption and can worsen liver overload.

**Fortified cereals and supplements** - many breakfast cereals, bars, and multivitamins contain added iron.

**Important note:** These changes only apply if you're at risk of iron overload due to **regular transfusions**.

For sickle cell warriors that don't have regular transfusions, eating more dietary iron can help to raise haemoglobin levels.

## Special situations: periods and hormones

If you have regular heavy periods, you naturally lose blood (and therefore iron) each month. This can sometimes protect against iron overload.

However, hormonal changes during your period cycle can also trigger pain and fatigue in people with sickle cell. For some, using hormonal contraception to stop periods may reduce these symptoms; but it also removes that natural iron loss. This balance is individual and its best discussed with your doctor or haematology team.

## Summary

Iron overload is a serious complication, but it can be managed. With regular monitoring and chelation therapy, iron levels can be kept under control, protecting your organs for the long term.

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# Crizanlizumab (Adakveo)

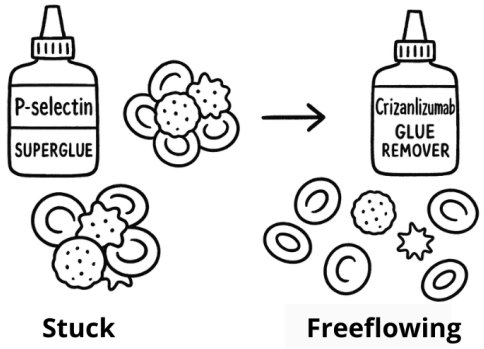
## What is it?

Crizanlizumab is a medicine designed to reduce sickle cell crises by stopping blood cells from sticking together and blocking small blood vessels.

P-selectin is a protein normally found on the surface of platelets and blood vessel walls. Its main job is to help the body form clots and start healing when a blood vessel is injured.

In sickle cell disease, though, P-selectin can become overactive. Instead of only helping platelets stick where they're needed, it also makes red blood cells, white blood cells, and platelets stick together and to vessel walls when they shouldn't. This clumping slows blood flow and can trigger a crisis.

Crizanlizumab works by blocking P-selectin. Think of P-selectin like superglue - it makes blood cells stick together when they shouldn't. Crizanlizumab acts like superglue remover, stopping the cells from clumping so blood flows more smoothly.



## How is it given?

Crizanlizumab is given as an **intravenous infusion (a drip into a vein)**.

- The first two doses are given 2 weeks apart, then it's usually once every 4 weeks.
- Each infusion takes about 30 minutes, though you may spend longer in clinic for monitoring.

Many patients report feeling **tired or achy for a few days after the infusion**, and sometimes they notice pain is worse just before their next dose is due.

## How effective is it?

The first major trial (*SUSTAIN*, 2017) showed promising results, with patients on the higher dose of crizanlizumab having around **45% fewer pain crises per year** compared to those not on it. Importantly, the benefit was seen **whether or not patients were already taking hydroxyurea**.

This led to it being licensed in the US and later in Europe.

However, later real-world studies and follow-up analyses didn't show as much benefit, especially compared with standard treatments like hydroxyurea. Results were mixed:

- Some patients found it reduced the number of crises.
- Others noticed little or no difference.

Because of this:

- In **Europe**, the European Medicines Agency (EMA) withdrew its licence in 2023.
- In the **UK**, NHS England and the Medicines and Healthcare products Regulatory Agency (MHRA) also reviewed the data and decided it wasn't effective enough to continue funding.

## Where is it used now?

**United States:** Crizanlizumab is still licensed and used, usually for patients who can't take or don't respond well to hydroxyurea.

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## Side effects

The good news is that crizanlizumab is generally safe, with no major long-term risks identified so far. Side effects tend to be mild and manageable:

- Nausea
- Joint pain (arthralgia) or back pain
- Fever
- Infusion-related reactions (such as chills, itching, or shortness of breath during the infusion - these are rare but monitored for in clinic)

Most people tolerate it well, though some report fatigue or increased pain around infusion times.

---

## Summary

Crizanlizumab is a safe medicine that aims to reduce sickle cell crises by stopping cells from sticking together. It showed promise in early trials, but later results were less convincing.

- In the **US**, it remains an option, particularly for people who cannot tolerate hydroxyurea.
- In the **UK and Europe**, it has been withdrawn because it wasn't effective enough to justify funding.

While it may help some people, it hasn't consistently shown the same level of benefit as other sickle cell treatments. The positive news is that crizanlizumab has a

good safety profile, so if more evidence emerges in the future, it may still have a role for some patients.

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## Section 3: Curative treatments

# Casgevy (Gene Therapy)

## What is it?

Casgevy is a breakthrough form of gene therapy with the potential to cure sickle cell disease. It uses a cutting-edge gene editing tool called **CRISPR**, which won its inventors the Nobel Prize in 2020.

Instead of just managing symptoms, Casgevy reprograms your own DNA so your body can permanently produce healthy red blood cells.

When I first heard about it in November 2023, it gave me so much hope. For the first time, it felt like real progress was being made towards a genuine cure.

Casgevy was approved for sickle cell disease in both the United States and Europe in 2023. In early 2025, it became licensed for people aged 12 and over with severe sickle cell and recurrent crises. It's not yet licensed for younger children, though research is ongoing. We're still a long way from it being accessible to the majority of people with sickle cell worldwide (it costs £1.6 million), but its approval marks a huge step forward.

## How does it work?

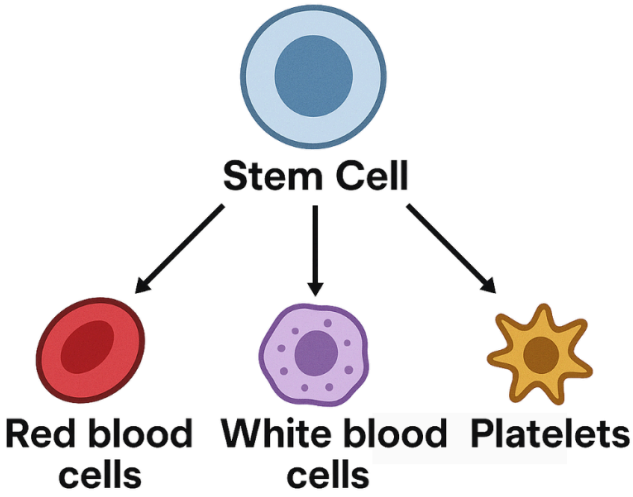
Casgevy works by switching back on **fetal haemoglobin (HbF)** the special type of haemoglobin we all make as babies. HbF is highly protective against sickling.

Normally, the body turns HbF off a few months after birth. Casgevy reprograms your blood-making system so HbF is turned back on permanently. That means new red blood cells are produced with HbF instead of sickle haemoglobin, protecting them from sickling.

**1)Collect Stem Cells from your bone marrow (2-3 months)**

Let's introduce another powerful player in the game of sickle cell treatment. The stem cells.

Stem cells are the “master cells” in your bone marrow that make all your blood cells - red cells, white cells, and platelets.

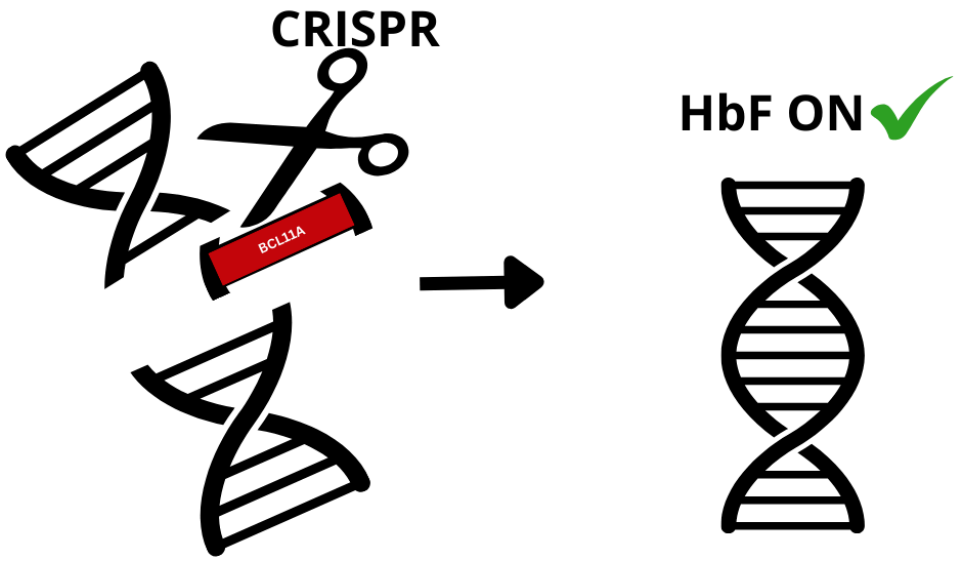



Stem cells are collected from your blood in a couple of sessions separated by 2 weeks. This ensures enough are collected and gives your body time to recover in between. Each session takes several hours, and medicines are given beforehand to help release stem cells from the bone marrow into the bloodstream. Then a process called apheresis is used:

- Blood is taken out through a vein in one arm.
- A machine spins the blood, separates out the stem cells, and returns the rest of the blood through the other arm.
- This usually takes a few hours, sometimes over two sessions.

**2) Treat Stem Cells with Casgevy (a few weeks, in the lab)**

This is where the gene editing magic happens. Scientists target a specific gene called **BCL11A**, which is responsible for switching off fetal haemoglobin. Casgevy uses CRISPR to cut out this gene, so your cells are permanently programmed to keep producing HbF.




**3) Chemotherapy (conditioning) (2-4 weeks)**

- Before the edited stem cells can be returned, your bone marrow has to be cleared to make space.
- This involves **high-dose chemotherapy**, usually given over 1-2 weeks, followed by time for your body to recover.
- Unfortunately this is where side effects can occur including: Hair loss, infections, fatigue, nausea, and risk of infertility (sometimes permanent).
- You'll usually stay in hospital during this phase.

**4) Reinfuse Casgevy-treated stem cells (1 day)**

- The edited cells are returned to you through a drip, similar to a transfusion.
- This part is quick, but you'll stay in hospital for close monitoring afterwards.

**5) Wait for recovery and engraftment (1–3 months)**

- It takes time for the edited stem cells to “engraft” (settle into the bone marrow and start producing blood cells).
- During this time your blood counts are low, so you’ll be closely monitored in hospital and then as an outpatient.
- Most patients start making healthy red cells with HbF within a few months.
- In studies, patients achieved around **50% fetal haemoglobin**, similar to the amount of normal HbA people who carry the sickle cell trait have, which is enough to eliminate symptoms.

From start to finish, the Casgevy process usually takes **around 6-12 months** including preparation, hospital stay, and recovery time.

## What does it achieve?

Clinical trials have shown dramatic results:

- Most patients had a **complete elimination of sickle cell crises**.
- HbF levels stayed high long after treatment.
- Many patients no longer needed any sickle cell medications afterwards.

In simple terms: Casgevy has functioned as a **permanent cure** in the patients treated so far.

## Risks and considerations

- **Short-term risks:** mostly from chemotherapy (hair loss, infections, infertility risk, sickness).
- **Long-term risks:** data are still being collected, but so far results are very encouraging with no major safety concerns.
- **Not suitable for everyone:** patients must be fit enough to tolerate chemotherapy and able to access specialised centres.

- Because Casgevy is so new, patients will need lifelong follow-up to monitor long-term effects.

## How is it different from bone marrow transplant?

Both treatments involve chemotherapy and reinfusing stem cells, but:

- **Bone marrow transplant** uses stem cells from a donor, so there is a risk of rejection or graft-versus-host disease.
- **Casgevy** uses your own stem cells, so there is no risk of rejection.

This makes Casgevy an exciting option for people who don't have a matched donor.

## Summary

Casgevy is one of the most exciting breakthroughs in sickle cell treatment. By using CRISPR to reprogram your own stem cells, it can permanently switch fetal haemoglobin back on and prevent red cells from sickling.

- Approved in the US, UK, and Europe for patients aged 12 and over with severe, recurrent crises.
- Still very new, with limited availability and high costs.
- Requires chemotherapy, so not everyone is eligible.

But for those able to access it, Casgevy has offered something we've all dreamed of: the chance to live free of sickle cell for life.

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# Bone marrow transplant (Stem cell transplant)

## What is it?

A bone marrow transplant, also called a **stem cell transplant**, is currently the only **established cure** for sickle cell disease.

It works by replacing your own sickle cell-producing stem cells with healthy donor stem cells. Once the donor's cells take over, your bone marrow produces red blood cells with **normal haemoglobin** instead of sickle haemoglobin.

Bone marrow transplants are most successful in **children**, especially when the donor is a **matched sibling**. Because the risks can be significant, doctors only offer it to people with severe sickle cell disease – for example, those with frequent painful crises, strokes, or organ damage.

## How does it work?

**1)Find a matched donor**

- Usually a brother or sister with the closest possible blood and tissue match.
- Unrelated or mismatched donors are possible, but outcomes are less predictable.

**2) Chemotherapy (conditioning)**

- You receive chemotherapy to destroy your own sickle cell stem cells and make space for the donor's cells.

**3)Infuse donor stem cells**

- The donor's healthy stem cells are given through a drip, similar to a blood transfusion.

**4)Engraftment**

- Over the next few weeks, the donor stem cells settle in the bone marrow and start producing normal red blood cells.

If all goes well, the patient's blood system is gradually replaced by the donor's, preventing sickling for life.

## Risks and side effects

**Short-term risks** (mainly from chemotherapy):

- Infections (due to low immunity while waiting for engraftment)
- Hair loss
- Fatigue
- Nausea
- Infertility risk (sometimes permanent)

**Long-term risks:**

- **Rejection:** The patient's immune system may destroy the donor stem cells, causing the transplant to fail.
- **Graft-versus-host disease (GVHD):** The donor's immune cells may attack the patient's tissues (skin, gut, or liver). This can range from mild to life-threatening and may require long-term immunosuppressive medicines.

Because of these risks, bone marrow transplant is not offered to every person with sickle cell.

## Success rates

Success rates depend heavily on the age of the patient and the type of donor match available:

- **Children with a matched sibling donor:** around **80–90% are cured**.
- **Adults:** outcomes are less predictable, with higher risks of complications.
- **Unrelated or mismatched donors:** success is possible but less reliable, and the risk of GVHD is higher.

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## How is it different from Casgevy (gene therapy)?

Both treatments involve chemotherapy and reinfusing stem cells, but the key difference is:

- **Bone marrow transplant** uses a donor's stem cells, which carries the risk of rejection and GVHD.
- **Casgevy** uses your own stem cells, edited in the lab, so there is **no risk of rejection** or graft-versus-host disease.

This is why Casgevy has created so much excitement - it may offer the benefits of transplant without some of the immune risks.

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## Summary

Bone marrow transplant is the only proven cure for sickle cell disease available today. It can be life-changing, especially for children with a matched sibling donor, with cure rates up to 90%.

However, the risks are significant, and most people with sickle cell do not have a matched donor. For this reason, transplants are reserved for the most severe cases.

With new treatments like Casgevy now approved, the future may hold safer and more widely accessible curative options - but for now, bone marrow transplant remains the original standard of cure.

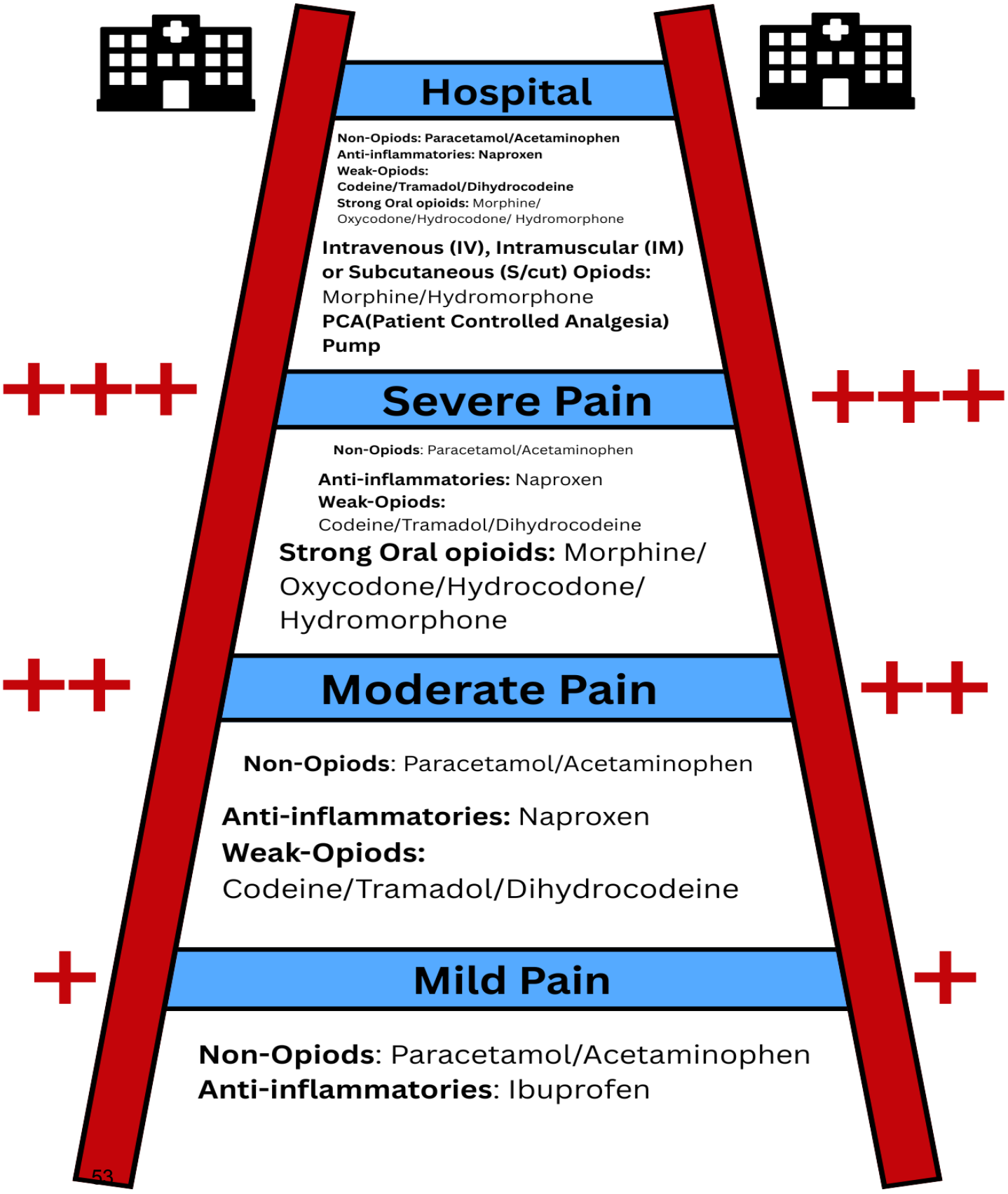
# Section 4: Pain medication

When I was a junior doctor deciding what specialty to choose, I seriously considered anaesthetics. As someone who has experienced severe pain myself, I thought it would be rewarding to have all the tools to relieve other people's suffering.

I didn't end up becoming an anaesthetist because I couldn't stand the nights and weekend work. But I learned a huge amount about pain management. Based on that clinical knowledge and my personal lived experience, I've created what I call **"The Sickle Cell Pain Ladder."**

This is an adaptation of the well-known **World Health Organisation (WHO) Pain Ladder**, which has guided doctors since 1986. My version is designed specifically with sickle cell crises in mind.

# The Sickle Cell Pain Ladder



People are sometimes more familiar with the brand names of some of these painkillers so I've added this below in the **Brand Names Table**.

| **Generic Name** | **Common Brand Names** |
|---|---|
| Paracetamol | Panadol, Calpol (children's), Disprol (UK) |
| Acetaminophen | Tylenol, Excedrin (US) |
| Ibuprofen | Nurofen, Advil, Motrin |
| Naproxen | Aleve (US), Naprosyn, Napratec |
| Codeine | Zapain, Co-codamol (with paracetamol), Tylenol#3(US, with acetaminophen) Solpadeine |
| Dihydrocodeine | DF118, Co-dydramol (with paracetamol), DHC Continus |
| Tramadol | Zydol, Ultram |
| Oral Morphine | Oramorph, Sevredol, MS Contin, Morphgesic |
| Oral Oxycodone | OxyContin (prolonged-release), Oxynorm (immediate-release), Percocet (with acetaminophen) |
| Oral Hydromorphone | Palladone (UK), Dilaudid (US) |
| Oral Hydrocodone | Vicodin (with acetaminophen), Norco, Lortab (US) |

## Step 1: Mild Pain

**Non-opioids:**

Paracetamol (also called Acetaminophen in the US/Canada).
On its own, it isn't very strong, but it works **synergistically** with stronger painkillers and provides more relief. It's always worth keeping it on board, even in severe pain. In hospital, IV paracetamol is sometimes given as it works better than tablets.

**Anti-inflammatories:**

Ibuprofen is the most common.

- Can irritate the stomach, so take with food.
- If used long-term, they can cause stomach ulcers.
- If you develop heartburn or stomach pain, you may need a protective anti-acid medicine such as omeprazole.

## Step 2: Moderate Pain

**Continue Step 1 medicines** (paracetamol + anti-inflammatory).

**Weak opioids:**

- Codeine
- Tramadol
- Dihydrocodeine

These are often used in combination with paracetamol.

## Step 3: Severe Pain

**Continue Step 1 + 2 medicines**.

**Strong oral opioids:**

- Morphine
- Oxycodone
- Hydrocodone
- Hydromorphone

At this stage, pain is often intense enough to interfere with daily function. Stronger opioids are necessary to get control.

## Step 4: Hospital Level Pain

If pain can't be controlled at home, hospital treatment is needed.

**IV, IM or Subcutaneous opioids:**

Morphine or Hydromorphone are most common. The choice of opioid varies by region and the individual patient.

For example in Nigeria IV Pentazocine is an opioid that's commonly used when Morphine is less available. Also Fentanyl is used for some patients resistant to other opioids.

**Patient-Controlled Analgesia (PCA):**

This is a pump that allows you to press a button to give yourself a safe dose of IV opioid when needed. This is the standard of care for uncontrolled sickle cell crisis pain.

## Managing Side Effects

All opioids, whether weak or strong, can cause side effects:

- **Drowsiness**
- **Nausea/vomiting** → request an anti-sickness tablet if needed.
- **Constipation** → if you take opioids regularly, you should also take a **laxative** to keep your bowels moving.

Side effects are manageable, and hospital teams are used to prescribing extra medicines to help. Don't be afraid to ask.

## Opioid Stigma and Reassurance

Accessing opioid medication is one of the most frustrating experiences for sickle cell warriors struggling with severe pain crises. A disease that's already so painful is made worse due to the stigma around opioids.

Because of the global opioid crisis, some healthcare professionals are overly fearful about prescribing them. And some sickle cell warriors are overly fearful about taking them. But here's what you need to know:

- Opioids are safe and effective when prescribed for severe pain.
- Using opioids for sickle cell crises is appropriate medical treatment, not abuse. Abuse means taking opioids inappropriately, for example when not in pain or at doses not prescribed.
- **People with sickle cell often need higher doses than others.** This reflects the severity of the pain and differences in tolerance, not misuse.

No one with sickle cell should feel judged or guilty for needing opioid pain relief.

If you ever feel like you may be abusing opioids then ask for help from your doctor or local drug rehab centre.

**Simple Opioid Conversion Guide**

| Opioid (Oral) | Approximate Equivalent to 10 mg Oral Morphine |
|---|---|
| **Codeine** | 100mg = 10 mg morphine (10x weaker) |
| **Dihydrocodeine** | 100mg = 10 mg morphine (10x weaker) |
| **Tramadol** | 100mg = 10 mg morphine (10x weaker) |
| **Hydrocodone** | 30 mg = 10 mg morphine (3x weaker) |
| **IV Pentazocine** | 30 mg = 10 mg morphine (3x weaker) |
| **Oxycodone** | 10 mg = 15 mg morphine (1.5x stronger) |
| **Hydromorphone** | 2 mg = 10 mg morphine (5x stronger) |
| **IV Fentanyl** | 0.1 mg (100 µg) = 10 mg morphine (100× stronger) |

Understanding these equivalents is useful if your doctor suggests trying a different opioid. It can help you consider the best dose to start with based on the strength of what you've taken safely before.

**For example:**

- 10 mg oral morphine = 100 mg codeine or 100 mg tramadol or 2 mg hydromorphone.
- If switching from oxycodone to morphine, you may need 1.5x the morphine dose.

**Helpful disclaimer:**

- These are approximate, **individual responses may vary**.
- Always follow the dosing advice from your healthcare provider.

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## Other Pain Medicines

Sometimes, additional "adjuvant" medicines are used, especially for long-term or nerve-related pain:

- Gabapentin or pregabalin
- Amitriptyline or duloxetine

These aren't typically used for acute crises but may help if you have chronic background pain.

---

## Summary

The Sickle Cell Pain Ladder shows you how pain relief is managed step by step, building from simple medicines through to stronger opioids and hospital treatments when needed.

The key things to remember are:

- No single medicine is usually enough. **Combinations work best.**
- **Opioids are not your enemy.** They're essential tools in controlling severe sickle cell pain.
- Side effects can be managed, so don't hesitate to ask for help with nausea, constipation, or drowsiness.
- The moment you're aware of being at stage 3 in the sickle cell pain ladder, prepare to go to hospital if your strong oral opioid home medications don't help.
- Sickle cell acute pain crisis is one of the most painful conditions known to medicine, you deserve effective treatment.

# Section 5: Vitamin supplements

## Vitamin Supplements and Sickle Cell

For a long time, I used to think supplements were a bit of a “cheat code”, a way of avoiding eating a healthy diet. My philosophy was that if you ate well, you shouldn’t need to take extra pills or powders.

But when I started digging into the research around nutrition in sickle cell disease, my perspective changed. I realised that even if you eat a healthy, balanced diet, what’s “enough” for most people might not be enough for sickle cell warriors.

Why? Because sickle cell places higher demands on the body than normal.

**Constant haemolysis:** Red blood cells break down far more quickly, so your body is constantly burning energy and nutrients to replace them.

**Oxidative stress and inflammation:** Both are higher in sickle cell, which increases the need for antioxidants and repair mechanisms.

**Reduced intake:** During a crisis, pain and fatigue often reduce appetite, and strong opioid medications can also blunt hunger or upset the stomach.

Put simply, sickle cell warriors live with increased nutritional demands while often taking in less. That creates a higher risk of vitamin and mineral deficiencies. Supplements aren’t a replacement for eating well, or medical treatment, but in sickle cell they can play an important supportive role. They provide your body with a consistent supply of nutrients that are difficult to maintain through diet alone. Over time, I came to see them less as a shortcut and more as a tool – one that can help keep my body more resilient against the unique pressures of sickle cell.

In this section, we’ll look at the key supplements shown to help in sickle cell: Vitamin D, Omega-3, Zinc and I’ll give my opinion on a few other special mentions that are less well studied.

# Vitamin D

Vitamin D supports bone health, immune function, and muscle strength. It's made in the skin through exposure to sunlight and is also absorbed from the diet.

## Why do you need it in sickle cell?

Vitamin D deficiency is common in the general population, but even more so in sickle cell because:

- The constant breakdown and replacement of red cells increases the demand for nutrients.
- People with darker skin (common in sickle cell) need more sunlight to make vitamin D.
- Fatigue, low mobility, or medical advice may reduce sun exposure.

Vitamin D controls how much calcium and phosphate the body absorbs – two key minerals for strong bones. For sickle cell warriors, this is especially important because:

1. Sickle crises often damage bone tissue.
2. Bone complications (like avascular necrosis) are common in SCD.
3. Research shows higher Vitamin D levels are linked to less bone pain and shorter hospital stays during crises.

Vitamin D also strengthens the immune system by helping infection-fighting cells work efficiently and by balancing inflammation, which is often higher than normal in sickle cell.

## Are there any side effects?

Vitamin D is very safe. Problems only occur with very high doses over a long time, which can cause:

- High calcium levels (hypercalcaemia),
- Nausea or kidney stones (rare).

## What dose is recommended?

- **General population:** 800-1,000 IU (20-25 micrograms) daily.
- **Sickle cell:** Most experts advise the same minimum dose, but taken consistently all year round. Depending on your blood levels, you may need a higher prescription only dose for a short period to boost them, your doctor can check this with a blood test and prescribe for you.

Vitamin D tablets are widely available in supermarkets and pharmacies.

**Parents' Note: Vitamin D and Children**

**Why it matters:** Children with sickle cell need vitamin D just as much as adults, maybe even more. Vitamin D deficiency in childhood can slow growth, weaken bones, and increase the risk of rickets (soft, deformed bones).

**Bone health:** Because sickle cell already increases the risk of bone complications, keeping vitamin D levels healthy is especially important to protect children's growing bones.

**Dosing:** The recommended dose for children is usually the same as adults (around 800-1,000 IU daily), but your child's doctor may advise testing vitamin D levels with a blood test and adjusting the dose if needed.

**Practical tip:** Vitamin D supplements for children often come as drops or chewable tablets, which can make them easier to take consistently.

# Omega-3 (Fish Oil, EPA & DHA)

When I first learned about omega-3's benefits in sickle cell, I was curious whether my own diet was giving me enough. I did a blood test to check my Omega-3 level and was shocked by how low it was. I realised I don't eat enough oily fish and probably never will, so I decided to start supplementing. Since then, I've been taking a high-strength omega-3 fish oil everyday. For me, it's reassuring to know I'm definitely getting enough to support my long-term health.

## What does it do?

Omega-3 is an essential fatty acid your body can't make by itself, so you have to get it from food or supplements. The two most useful forms are EPA and DHA, mainly found in oily fish and algae. Plant sources like nuts and seeds contain ALA, but the body only converts about 5% into EPA/DHA.

In sickle cell disease, omega-3 helps by:

- Reducing inflammation in the blood,
- Making red blood cells more flexible,
- Lowering the stickiness of blood cells, reducing blockages.

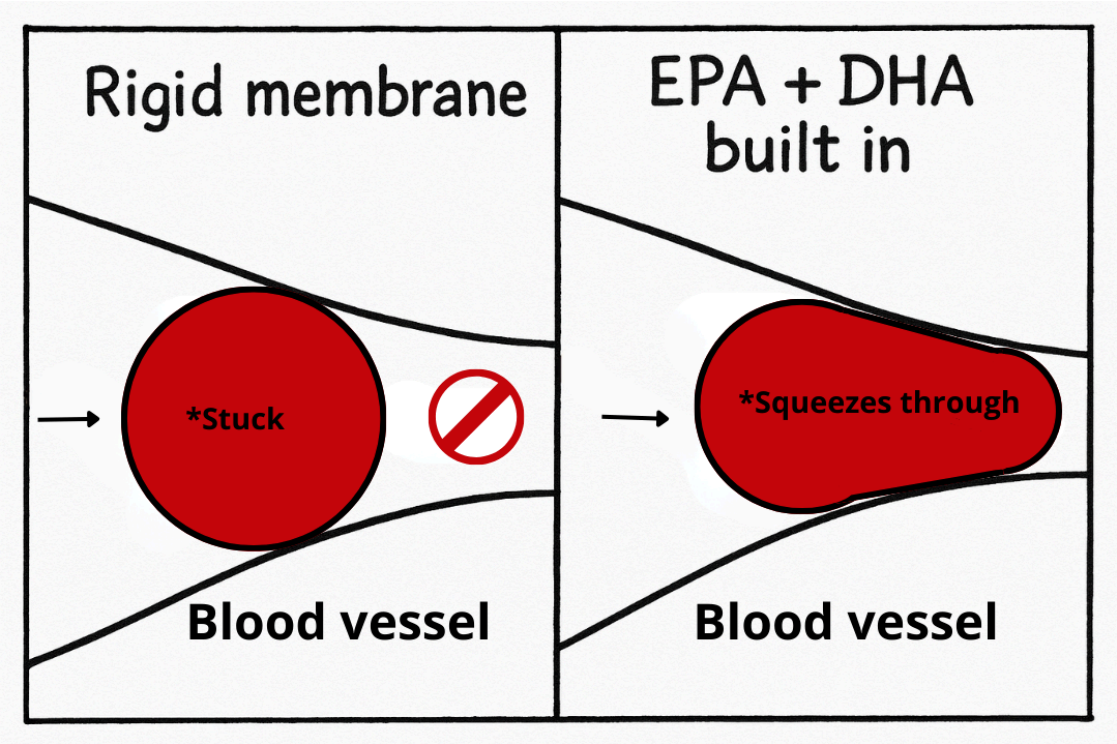
## Why do you need it?

We need to restore a healthy balance between **Omega-3** and its counterpart **Omega-6**. Omega-6 is another essential fatty acid in the body which drives inflammation and clotting. Research has shown that people with sickle cell often have too much Omega-6 compared to Omega-3. A normal healthy balance would be a **ratio of 3:1**, that's 3 times as much Omega-6 compared to Omega-3. Most people even without sickle cell have much higher ratios. Mine was 43:1 when I checked recently prior to starting supplements.

Restoring a healthy balance by increasing Omega 3 levels has been found in some studies to lower the number of sickle cell crises and shorten hospital stays.

## How does it work?

EPA and DHA get built into the red cell membrane. This makes the cells less rigid and less likely to sickle when passing through small vessels. At the same time, omega-3 reduces inflammation and clot formation in the bloodstream.




## What dose is recommended?

Research suggests 2-3 grams per day of combined EPA + DHA is needed for benefit in sickle cell. Most supermarket capsules only contain ~200-300 mg each, which means you'd need 8-10 per day to reach the target. High-strength supplements are available and more practical, though more expensive.

## Are there any side effects?

Omega-3 is generally safe. Possible mild effects include:

- Fishy aftertaste, nausea, or diarrhoea.
- High doses may slightly increase bleeding risk in people taking blood thinners.

**Parents' Note: Omega-3 and Children**

Omega-3 supports growth, brain development, immune balance, and may reduce pain crises in children with sickle cell.

Typical research doses are around **1-2 g/day** of combined EPA + DHA (about **15-20 mg per kg** body weight). Always check with your child's haematology team before starting.

## Why Diet Matters as well

**Omega-3** has to compete with its rival **Omega-6** for absorption. Omega-6 levels are high in processed foods and some nuts, especially Brazil and Cashews. Look at the tables below to see how you can change up your diet to reduce Omega-6. One of the main changes I made to my diet was to swap out the Brazil nuts I have in my breakfast smoothie for Walnuts and Flaxseed. Take a look at the tables below to start considering how you could change your diet to **decrease Omega-6** and **increase Omega-3.**

## Omega-3 Foods ⬆️

| Food | Notes |
|---|---|
| Oily Fish (Salmon, Mackerel, Sardines, Herring, Trout) | Best natural source of EPA and DHA |
| Algae Oil (Vegan) | Plant based source of DHA good alternative to fish |
| Walnuts | One of the few nuts high in omega-3 (ALA form) |
| Chia seeds | High in ALA, sprinkle into smoothies/oats |
| Flaxseed ground or oil | Rich in ALA, needs conversion into EPA/DHA |
| Rapeseed(canola oil) | Contains some ALA better balance than sunflower/corn oil |

## Omega-6 Foods ⬇️

| Food | Notes |
|---|---|
| Sunflower oil | Very high in Omega-6 common in processed foods |
| Corn oil | High in Omega-6 avoid for better balance |
| Soybean oil | High in Omega-6 found in processed foods |
| Brazil nuts | Healthy but high Omega-6: Omega-3 ratio |
| Cashews, Peanuts, Almonds | Generally low Omega-3, high in Omega-6 |

## Supplements

| | |
|---|---|
| **High quality Fish oil** | Look for IFOS-certified, ~2–3 g EPA+DHA/day |
| **Algae Oil Capsules** | Vegan-friendly DHA, good for fish allergies |
| **Krill Oil** | Contains EPA+DHA + astaxanthin antioxidant. Only low dose capsules available |
| **Combination formulas with Vitamin D** | Helpful as Vitamin D deficiency common in SCD |

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# Zinc

Zinc is an essential mineral involved in hundreds of processes in the body. It supports growth, immune defence, wound healing, and helps reduce damage from oxidative stress. In sickle cell, research suggests zinc can:

- Improve growth in children,
- Speed up wound healing, especially leg ulcers,
- Reduce the frequency of infections,
- Lower the risk of painful crises.

Most studies so far have been done in children, but the same benefits are thought to apply in adults too.

## Why do you need it?

People with sickle cell are more likely to have zinc deficiency because:

- Zinc is lost in higher amounts in the urine,
- The body has higher nutritional demands due to constant red cell breakdown,
- It can be hard to get enough from diet alone.

Low zinc levels are linked to delayed growth, weaker immunity, leg ulcers, and delayed puberty/sexual maturation.

## How does it work?

Zinc helps repair damaged cells, make new DNA, and strengthen immune cells. For sickle cell warriors, this means:

- Red blood cells recover better from daily damage

- Leg ulcers heal more quickly
- The immune system is stronger to fight infections
- Children and teenagers are more likely to have normal puberty

## What dose is recommended?

- General daily requirement: Adults ~8-11 mg/day, children ~5-10 mg/day.
- In sickle cell, some studies used 10-25 mg/day to correct deficiency.

Higher doses should only be taken under medical supervision. If you have poor healing leg ulcers or other signs of deficiency, ask your doctor to check your zinc levels and advise on dosing.

## Are there any side effects?

Zinc is safe at recommended doses. High doses can cause nausea, diarrhoea, or stomach pain. Very high intake over time can interfere with copper absorption, leading to deficiency.

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## Other Nutrients Being Studied (Special Mentions)

If you've read this far, you'll have noticed a common thread running through most sickle cell treatments and supplements: they aim to protect red blood cells from being damaged and broken down too quickly. Whether it's through reducing oxidative stress, improving flexibility, or supporting repair processes, the same pathways keep coming up.

The nutrients below don't yet have the same level of evidence as Vitamin D, Omega-3, Zinc, or Folic Acid (and L-glutamine, which we covered earlier in the Endari section). But they've shown some promise in small studies, and they may play a supportive role in keeping red cells healthier for longer.

### Vitamin E

A powerful antioxidant. Small studies suggest it may reduce red blood cell breakdown and improve their survival. Not enough evidence to recommend universally, but it may support general antioxidant defence.

### Vitamin C

Also an antioxidant. It supports immune function and helps protect red cells from oxidative damage. Evidence in sickle cell is limited, but it may have a role in overall blood health.

### Magnesium

Helps red blood cells stay hydrated. Some studies suggest supplementation may reduce the tendency of red cells to sickle by preventing dehydration, but results have been mixed.

### L-Carnitine

An amino acid derivative involved in energy metabolism. Early research suggests it

may reduce oxidative stress and haemolysis, but studies are still small and experimental.

## Summary

These nutrients are **likely to be helpful** for sickle cell warriors based on their properties, and I'm personally open to taking them. But at the moment, there isn't enough scientific evidence for me to go into detail on them.

They are, of course, all beneficial for general health even without sickle cell, which is why you'll often find them included in multivitamins. The unanswered question is what dose - if any - produces a significant benefit specifically in sickle cell disease.

If you're curious about them, it's best to discuss with your doctor before starting, especially if you're already taking other medications or supplements.

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## Bonus section: Ciklavit

Nigeria has the highest number of people living with sickle cell in the world. Many of my Nigerian followers reached out to tell me about a supplement called Ciklavit, which they've found helpful.

Because of this, I wanted to give it special attention here as a *bonus.* Ciklavit is marketed and widely used in Nigeria and parts of West Africa, so if you're reading this from that region, you've probably heard of it already.

While the scientific evidence is still limited, and it's not a substitute for medical treatment, I think it's important to acknowledge and explore it - especially since many sickle cell warriors in Nigeria have personal experience with it.

Ciklavit reportedly helps to reduce the frequency and severity of painful crises and improve overall quality of life for sickle cell warriors.

### How does it work?

Ciklavit is marketed as a supportive therapy to:

- Boost antioxidant levels
- Improve blood health
- Support energy levels

Sickle cell warriors live with constant red blood cell breakdown, much of it driven by oxidative stress. **Antioxidants** help protect red cells from this damage so they survive longer and energy levels increase. Ciklavit contains **Cajanus cajan (pigeon pea) extract.** Some small Nigerian trials suggested this may mildly increase HbF levels, but these results haven't been widely replicated.

### What's inside?

As well as the *Cajanus cajan* (pigeon pea) extract it contains an additional blend of vitamins and minerals. The typical formula includes:

- Vitamin C (ascorbic acid)
- Vitamin B-complex (B1, B2, B3, B5, B6, B7, B9, B12)
- Vitamin D
- Vitamin E
- Zinc
- Selenium
- Magnesium
- Calcium
- Essential amino acids (including phenylalanine, thought to have anti-sickling properties)

Exact amounts may vary between products and manufacturers so always check the product label.

## What dose is recommended?

Ciklavit is available as a syrup or capsule. The typical marketed dose is:
**Adults:** ~10 ml of syrup or two capsules twice daily,
**Children:** 5-10 ml of syrup twice daily.

⚠️ Important: These are manufacturer's recommendations, not international guidelines. There are no large-scale clinical trials to confirm optimal dosing. If you choose to use it, always follow the product instructions and discuss it with your doctor.

## Are there any side effects?

Ciklavit is generally well tolerated. Reported side effects are mild and uncommon, such as:

- Nausea, bloating, diarrhoea,
- Allergic reactions in sensitive individuals.

## Summary

While some small studies and case reports from Nigeria suggest benefits, there are no large-scale international clinical trials to confirm its effectiveness. Most of the evidence comes from testimonies rather than robust research.

If it’s available in your area, it may be worth trying, as no major side effects have been reported. The key is to monitor how you feel and decide whether it benefits you personally.

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# Sickle Cell Treatments Summarised

⚠️ *Always consult your haematologist before starting or changing any treatment.*

| ✅The Essentials (Safe & Proven) | |
|---|---|
| Folic Acid | → Supports red cell production; low risk, widely recommended. |
| Penicillin (children <5) | → Prevents life-threatening infections; very safe |
| Hydroxyurea (Hydroxycarbamide) | → Gold standard; reduces crises, stroke risk, and organ damage; decades of proven benefit . |
| Blood Transfusions (simple & exchange) | → Lifesaving in acute crises, stroke prevention; risks include iron overload . |

| ➕Helpful Extras (Good for Many People) | |
|---|---|
| Endari (L-glutamine) | → Antioxidant support; reduces crises in some, esp. if hydroxyurea not tolerated. |
| Vitamin D | → Deficiency common in warriors; supports bone and immune health |
| Omega-3 (EPA/DHA) | → May reduce sickling and inflammation |
| Zinc | → Supports immunity and growth; reduced infections/leg ulcers evidenced |
| Other nutrients (Vitamin C, E, Magnesium, L-Carnitine) | → Under study, may help but less robust evidence. |

| 🚧 New & Experimental (Promising but Limited Access) | |
|---|---|
| Crizanlizumab (Adakveo) | → Reduced crises by blocking cell "stickiness," but withdrawn from NHS use |
| Casgevy (Gene Therapy) | → First approved curative option; very limited access, long-term outcomes still under study . |

| **Bone Marrow / Stem Cell Transplant** |
|---|
| Oldest established cure for sickle cell. Best outcomes in children; risky, requires a matched donor . |

| **Regional/Traditional** |
|---|
| Ciklavit (Nigeria/West Africa) → Herbal supplement with antioxidant blend; widely used, but little large-scale clinical data . |

## Key Takeaway

If you remember one thing:

- Hydroxyurea is the best treatment – safe, proven, and effective for most.
- Support it with folic acid, vitamin D, omega-3, and zinc.
- Transfusions and antibiotics remain essential when needed.
- Newer drugs and gene therapies are exciting but not widely available yet.

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# Conclusion

In March 2025 I was admitted to hospital with a sickle cell crisis for the first time in 10 years. It was a surreal experience to be back in an environment that had caused me so much pain as both a patient and a doctor.

What struck me most was how little had really changed. I expected to just sit back and focus on recovering, confident that I'd be cared for without judgement or ignorance. Surely, I thought, I'd receive the same standard of care I always gave to patients in crisis. But no, I still had to fight. I had to use my medical knowledge and authority to receive appropriate treatment. And it made me angry to think of how many other sickle cell warriors are out there having to accept inappropriate treatment due to lack of knowledge.

I don't share this to pit warriors against healthcare staff. I think that until sickle cell gets the awareness it deserves that conflict will always be there. And then best way to resolve it is with knowledge. It's knowing your condition, understanding your options, and feeling confident in the steps you need to take to stay as healthy as possible.

That's why I wrote this book, to give you the tools to make confident treatment decisions and to better understand what's happening in your own body.
Life with sickle cell brings challenges and unpredictability that none of us can fully control. But there are things we *can* control. Our mindset. The way we prepare. And the steps we take to give ourselves the best chance of living well.

Through writing this book, I've learned a lot myself. My curiosity as both a sickle cell warrior and doctor drove me to keep asking questions: How do medications actually work? Why do side effects happen? And how can supplements actually help? My goal has been to present that understanding in a way that's clear and practical, without overwhelming you with unnecessary details at this stage.

Together, we've explored the treatments that form the backbone of sickle cell care -from folic acid, penicillin, and hydroxyurea, to transfusions, Endari, Casgevy, and Stem Cell transplant. We've also looked at supplements like Vitamin D, Omega-3, and Zinc, which can give your body extra support, and touched on emerging supplement considerations and region-specific options such as Ciklavit.

What I've shared here is my best attempt to pull together the most up-to-date understanding of sickle cell treatments and supplements available today. Research is moving, and I hope new options will continue to emerge. But right now, this is the clearest picture I can give you - the essential things you need to know to support your health without getting lost in confusion.

We live in a world overflowing with misinformation. The more I've seen it causing unnecessary stress and anxiety to people the more I've realised the need to speak up. I know the value of clear, honest information when you're trying to make sense of a condition like this. My life experience has put me in the perfect position to help.

So my hope is that the pages you've just read give you not only clarity, but also confidence - confidence to ask the right questions, to trust what truly helps, and to take control where you can.

Thank you for reading.

Stay Healthy,

Lewis

# Notes

1) Hydroxyurea Therapy in Sickle Cell Disease Multicenter Study of Hydroxyurea in Sickle Cell Anemia (MSH Study) EFFECT OF HYDROXYUREA ON THE FREQUENCY OF PAINFUL CRISES IN SICKLE CELL ANEMIACharache, et al.. NEJM, 1995

2) Thankachan, P., Walczyk, T., Muthayya, S., Kurpad, A. V., & Hurrell, R. F. (2008). Iron absorption in young Indian women: the interaction of iron status with the influence of tea and ascorbic acid. *The American Journal of Clinical Nutrition*, 87(4), 881–886. https://doi.org/10.1093/ajcn/87.4.881

3) von Siebenthal HK, Zimmermann MB, Hurrell RF. *Effect of dietary factors and time of day on iron absorption.* American Journal of Heamatology. 2023. In this, the authors note that adding a glass of orange juice to mixed meals increased iron absorption by approximately 2–3-fold.

# Further Reading & Resources

If you'd like to explore more about sickle cell treatments and support, here are some trusted resources:

- **Sickle Cell Society (UK)** – www.sicklecellsociety.org
  Leading UK charity offering information, advocacy, and patient support.

- **NHS – Sickle Cell Disease Overview** –
  www.nhs.uk/conditions/sickle-cell-disease
  Clear guidance on symptoms, treatments, and living with sickle cell.

- **CDC – Sickle Cell Disease Information (US)** –
  www.cdc.gov/ncbddd/sicklecell
  US-based resource with facts on complications, prevention, and care.

- **NIH: National Heart, Lung, and Blood Institute** –
  www.nhlbi.nih.gov/health-topics/sickle-cell-disease
  Evidence-based information and updates on treatments and research.

# About the Author

**Dr. Lewis Thomas, MBBS, MRCGP**
*Author, Educator, Sickle Cell Warrior*

Lewis Thomas is both medically trained and personally experienced in living with sickle cell disease. He studied medicine at the University of Nottingham, completing 10 years of medical training from medical school through postgraduate training, before going on to practise for 5 years as a fully qualified GP (family doctor) in Manchester, UK.

As a doctor, he learned how important it is to communicate clearly, taking time to explain treatments in simple, relatable terms. As a sickle cell warrior, he understands first-hand the challenges, fears, and unanswered questions that come with managing the condition.

Today, Lewis dedicates his work to empowering people with sickle cell through education and coaching. His mission is to provide trustworthy, accessible knowledge so that every warrior feels confident in their treatment decisions, prepared for conversations with healthcare professionals, and supported in living the healthiest, fullest life possible.

When he's not writing or coaching, Lewis shares educational content across social media to raise awareness, challenge stigma, and inspire warriors to take control of their health.

**To continue learning and receive ongoing clarity you can trust, please see the final page of this ebook for details on how to join Lewis's email community.**

# Stay Informed. Stay Empowered. Stay Healthy

If this guide has helped you feel clearer and more confident about sickle cell treatments, you don't have to stop here.

I share insightful and practical tips on how to stay healthy with sickle cell disease, physically, mentally and spiritually.

By joining my email list, you'll:

> ✅ Stay up-to-date on the **latest medicines, therapies, and supplements** relevant to sickle cell.
>
> ✅ Get **exclusive insights and resources** I only share with my email community.
>
> ✅ Be the first to know about **future guides, tools, and support programmes** designed to make your life easier.

📌 **Join today and get ongoing clarity and confidence for your sickle cell journey.**

👉 **Join here**