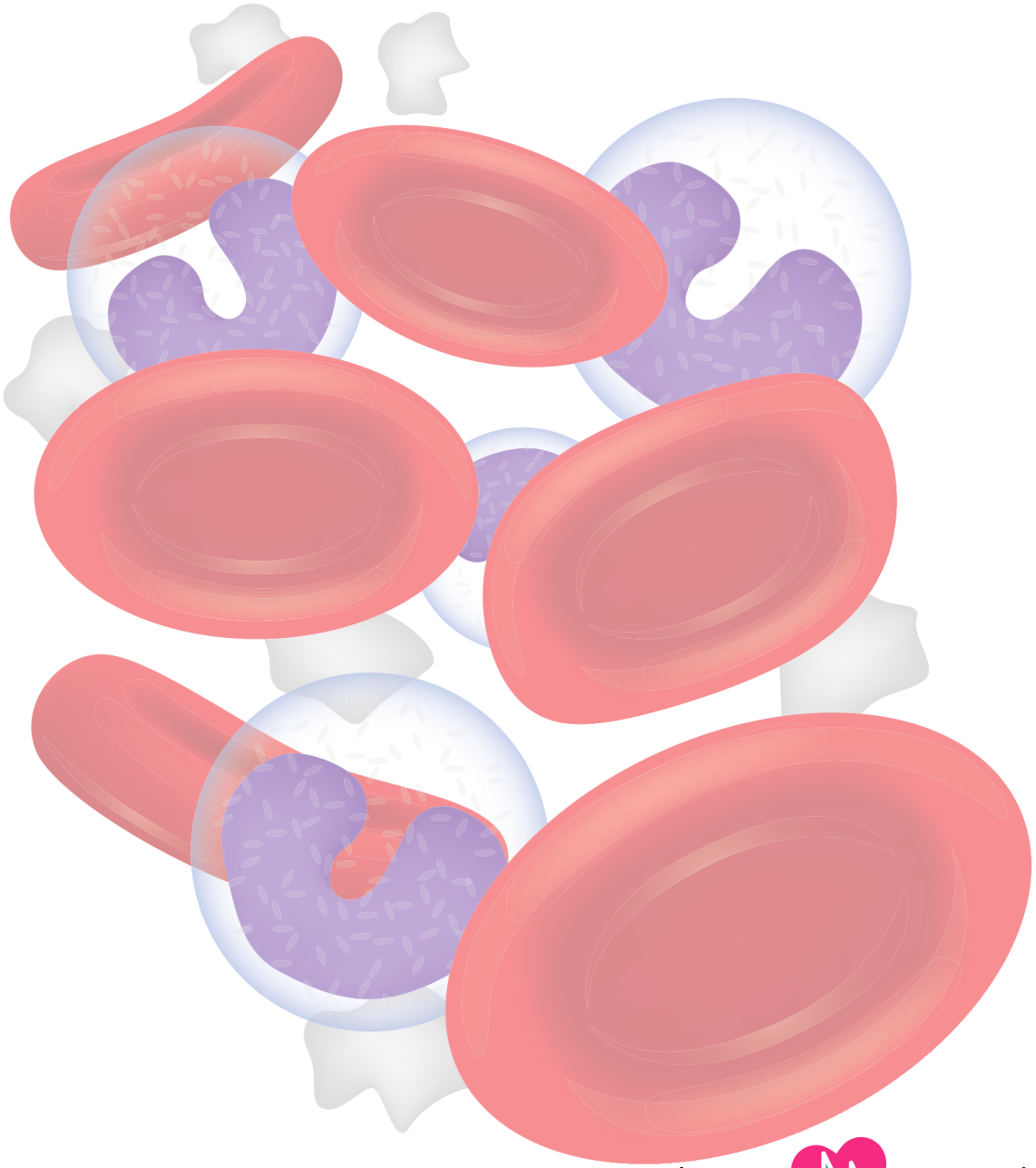
STUDY GUIDE

SICKLE CELL ANEMIA



Immunology NursingSOS

STUDY GUIDE

DEFINITION

Sickle cell anemia happens when the hemoglobin part of the red blood cell is misshapen. There are millions of hemoglobin molecules inside each red blood cell, and during sickle cell anemia, some of these hemoglobin molecules are abnormal and misshapen. This causes the red blood cells to form a sickle shape, clump together, and lose their elasticity. There ends up being clumps of sickled cells floating around the body, and this leads to many complications.

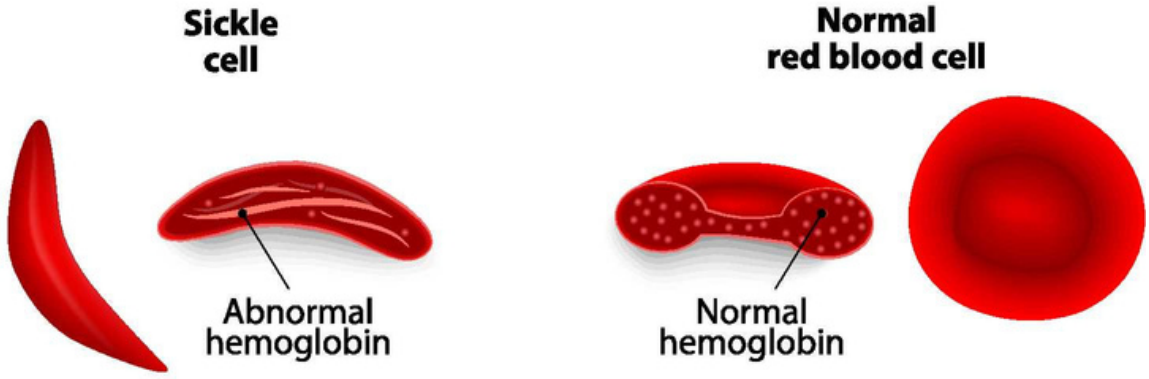


Image of abnormal hemoglobin comparison
designua / stock.adobe.com

CAUSES

Sickle cell anemia is an autosomal recessive condition. The patient would need to inherit one abnormal gene from their mother and one abnormal gene from their father. If they only receive one gene from either the mother or father, then they would be a carrier for sickle cell anemia and wouldn't have many symptoms themselves. However, they could pass it onto their children.

PATHOPHYSIOLOGY

Step 1: The Sickle Cell Trait is Inherited

The first step is that the trait for sickle cell disease is inherited. It is an autosomal recessive condition and the abnormal gene would have to be inherited from both parents. If the patient only receives one gene from either the mother or father, they would be a carrier for sickle cell anemia and wouldn't have many symptoms themselves.

Step 2: The Red Blood Cells Lose Their Oxygen & Causes Mis-shapen Hemoglobin

Step 2 happens as the red blood cells are oxygenated and deoxygenated. Normally, red blood cells go to the lungs and oxygen binds to hemoglobin molecules on those red blood cells. The red blood cells travel around and give that oxygen to the tissues throughout the body and then go back to the lungs to be re-oxygenated.

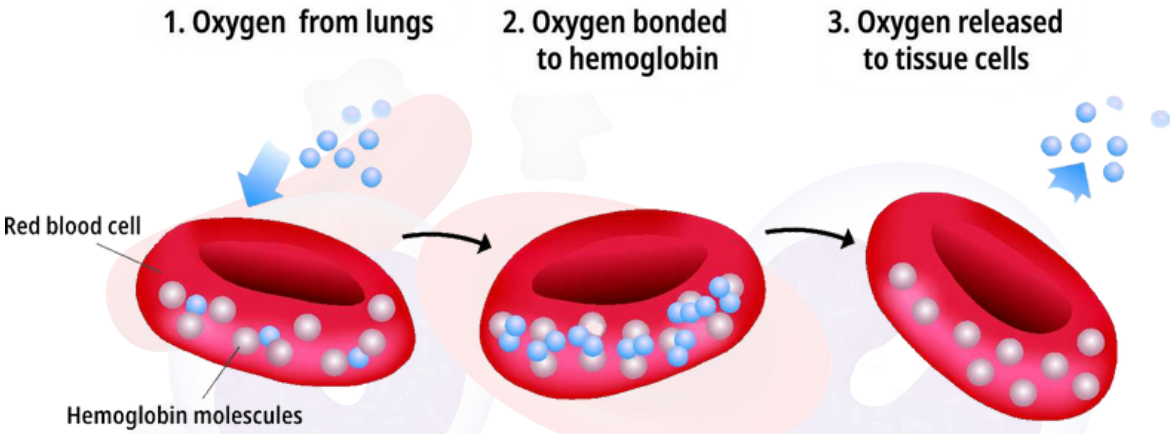


Image of hemoglobin carrying oxygen
Akarat Phasura / stock.adobe.com

However, during sickle cell anemia, when the hemoglobin on the red blood cells give up their oxygen, they start to misshape, like the oxygen molecules were helping the hemoglobin keep their shape; without it, the hemoglobin becomes misshapen.

If the cell is able to get oxygen again, it can un-sickle itself, but the more and more it sickles, the more damaged it becomes and the more it won't be able to go back to its normal shape.

> *Sickling of red blood cells is worse when oxygen demand increases, like during a fever, dehydration, or stress. These are potential triggers for a sickle cell crisis.*

Step 3: Hemoglobin Molecules Clump Together & the Red Blood Cell Sickles

When the hemoglobin are deformed like this, they start to clump together inside the red blood cells and form long strands of abnormal hemoglobin. This is what causes the red blood cell to form a sickle, or crescent, shape.

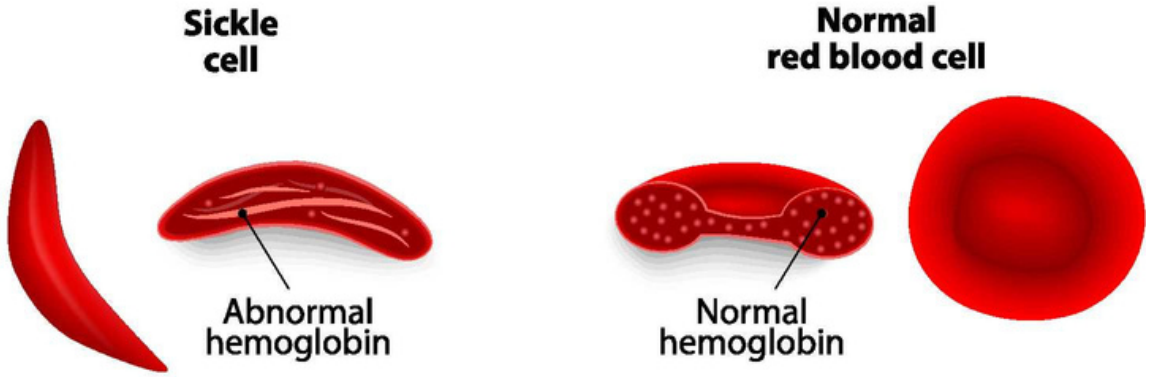


Image of abnormal hemoglobin comparison
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Step 4: Complications

When this happens and the red blood cells form a sickle shape, they aren't able to carry oxygen as well as healthy red blood cells, so they can't take as much oxygen to the tissues. They also start to clump together and get stuck in small blood vessels, which can:

- Reduce blood flow to those tissues
- Lead to stroke
- Block vessels in the lungs and kidneys

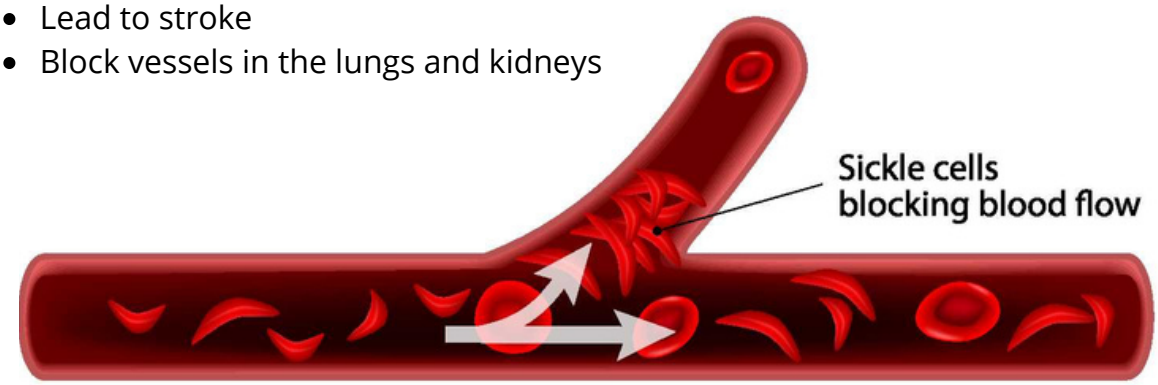

Image of sickle cells blocking blood flow
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Step 5: Sickled Blood Cells are Destroyed

As the red blood cells become sickled, they become weaker and weaker and get destroyed a lot faster than normal red blood cells. Normal red blood cells last between 90 to 120 days in the body, but cells that are sickled only last about 10 to 20 days. They have a much shorter lifespan.

There are many complications of this quick destruction of red blood cells, like anemia and the bones needing to work on overdrive to produce more and more red blood cells.

SIGNS AND SYMPTOMS

There are 2 underlying causes for most signs and symptoms of sickle cell anemia:

- The sickled cells clump together in the blood vessels and block blood flow to the tissues
- The cells are destroyed at a much faster rate than healthy red blood cells

These symptoms are related to the lack of normal (unsickled) RBC, and the impact of lack of oxygen and blood flow.

Pain

Pain is one of the biggest initial and ongoing symptoms associated with sickle cell disease. There is pain because the sickled red blood cells are clumping together and blocking the blood flow to specific areas of the body, like the fingers, toes, and joints.

This can be noticed in infants usually 5-7 months old, when they are usually first diagnosed. They may have:

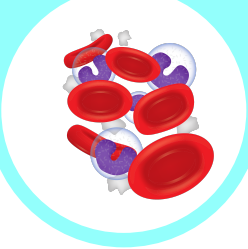
- Swollen hands and feet
- Increased fussiness
- A fever

The pain can be acute, which is called a pain crisis or a vaso-occlusive crisis, and last for a few hours to days, or it can be chronic from long term damage of blocked of blood flow to bones, GI tract and joints, causing bone pain, ulcers, or joint pain.

- **Pain Crisis / Vaso-Occlusive Crisis**: Sickled cells clump together and block blood flow
- Symptoms:
  - Fever
  - Pain
  - Swelling

Swollen/Inflamed Fingers & Toes

Due to the lack of blood flow from the blockage of sickled red blood cells, the fingers and toes can be swollen and inflamed, which is called dactylitis.



Anemia, Tiredness, Tachycardia, & Pale Skin

Anemia is a huge symptom that can cause tiredness, tachycardia, and pale skin. Anemia means that the red blood cell count is low; the sickled red blood cells are much more fragile than typical healthy red blood cells:

- **Healthy RBCs**: Last 90-120 days
- **Sickle Cell RBCs**: Last 10-20 days and then burst, leaving the body trying to constantly keep up and create more red blood cells

This causes anemia, and with fewer healthy red blood cells to carry oxygenated blood to the tissues and organs. The heart will try to compensate by increasing the heart rate to get more and more blood to the body, leading to tachycardia. The overall tiredness is due to the lack of blood flow to keep up with the body's metabolism and exertion.

Stunt Growth

If the lack of blood flow is chronic, it can cause a delay or stunt in growth and puberty. The body is focusing on trying to keep up with making healthy red blood cells to oxygenate properly, and body growth gets put on the back burner while the body oxygenates vital organs.

Increased Risk of Infection

Increased risk of infection is another symptom and complication of this condition. The patient might be constantly getting sick; the body has to process all these dead sickled red blood cells at a much higher rate since they don't last as long, so the organs that process red blood cells, like the spleen, can get backed up and become damaged over time. With the spleen focusing on filtering the sickled RBC out, it can't help as much with the immune response to help the body fight off infections.

The blood cells within the spleen may also get blocked with those clumps of sickled cells, so the spleen won't be able to function as well since it's not getting as much blood and oxygen. This can lead to a condition called:

- **Splenic Sequestration**: Blood pools in the spleen and it becomes very enlarged
- Symptoms:
  - Severe anemia
  - Difficulty breathing
  - Tachycardia
  - Hypovolemic shock

When this happens suddenly, it is called acute splenic sequestration and is a medical emergency. The patient will be very anemic, have trouble breathing, have tachycardia, and can go into shock very quickly. All of that blood and fluid is just sitting in their spleen and not going to their other organs.

Splenic sequestration occurs when sickled cells block blood flow in the spleen. This leads to massive swelling of the spleen and spleen enlargement. It leads to severe anemia, difficulty breathing, tachycardia, and shock. It is a medical emergency and should be treated immediately.

Since the lifespan of the sickled cells is so much lower than healthy red blood cells, if there are many that are sickled and destroyed at the same time, like during a fever or times of stress, it can lead to:

- **Hyperhemolytic Crisis**: Large amount of red blood cells rupturing at once
- Symptoms:
  - Anemia
  - Jaundice
  - Reticulocytosis

This can lead to anemia, since all of those blood cells are being destroyed, and jaundice. Normally, when red blood cells are destroyed, the hemoglobin is made into bilirubin, which is a yellowish colored substance that is normally processed by the liver. However, with so many red blood cells being destroyed all at once, the liver isn't able to process the bilirubin fast enough, so it stays in the body, leading to jaundice, which is a yellow tint of the skin and sclera of the eyes.

The bone marrow will also start producing a lot of baby red blood cells that aren't able to function as well as mature red blood cells. This is called reticulocytosis and can happen during a hyperhemolytic crisis since the bone marrow is trying to make up for all of the red blood cells that are being destroyed, but is just making baby red blood cells.

If red blood cells are being destroyed faster than the bone marrow can produce normal healthy ones, then the patient could go into another type of crisis:

- **Aplastic Crisis**: There is not enough red blood cells in the body to keep the organs perfused and oxygenated
- Symptoms:
  - Severe anemia

An aplastic crisis occurs when red blood cells are destroyed and not enough are being made to replace them. There aren't enough red blood cells to keep the organs and tissues perfused and they aren't getting enough blood and oxygen that they need to function. This leads to severe anemia and is a medical emergency!

NURSING ASSESSMENT

There are 2 underlying causes for most nursing assessments of sickle cell anemia:

- The sickled cells clump together in the blood vessels and block blood flow to the tissues
- The cells are destroyed at a much faster rate than healthy red blood cells

Pain Level

Monitor your patient's pain level and make sure their pain is controlled by either oral or IV pain medications. The pain can be chronic and acute and the chronic pain can be exacerbated by an acute pain crisis as well. Sickle cell anemia can be very painful as those clusters of abnormal red blood cells get stuck in the blood vessels. This can lead to a lack of blood flow to the tissues, causing a lot of pain.

Respiratory Status

Keep a close eye on their respiratory status to make sure their lungs are clear and they are breathing comfortably. If the sickled cells clump together and get lodged in the lungs, it can lead to respiratory distress. Acute Chest Syndrome is a common respiratory condition that can happen with sickle cell disease where the sickled cells block blood flow to the lungs and the lungs become inflamed, leading to hypoxia.

Be sure to watch for:

- A change in breathing pattern

- Fever
- Low oxygen saturations (less than 90%)
- Cough
- Chest pain

If sickled cells clump together and block blood flow within the lungs, it can lead to acute chest syndrome. This is a medical emergency and must be treated immediately to help restore oxygen to the body.

Vital Signs

Assess their other vital signs:

- Temperature
- Heart rate
- Respiratory rate
- Blood pressure
- Oxygen saturation

Make sure their organs are being perfused properly. Any changes in these vital signs may indicate a sickle cell crisis, like vaso-occlusive crisis or splenic sequestration.

Hematocrit & Hemoglobin

Monitor their hematocrit and hemoglobin levels to make sure those are staying stable. You absolutely need to keep an eye out for a sudden drop in their H&H, which could indicate splenic sequestration, hyperhemolytic crisis, or aplastic crisis. They may need a blood transfusion to keep their tissues perfused.

Monitor for Gallstones

Monitor the patient for symptoms of gallstones:

- Jaundice: A yellowing of the skin and the sclera of the eyes and is caused by an increase in bilirubin
- Nausea and vomiting
- Severe abdominal pain

Since so many sickled RBC are being destroyed it will increase billirubin.

These patients are at an increased risk of gallstones because there are so many sickled cells that are being destroyed, causing an increase in bilirubin. A chronic increase in bilirubin can lead to gallstones because the gallbladder cannot keep up with the filtering process.

Neurological Changes

Watch for neurological changes; these patients are at higher risk of stroke due to the shape and clumping of the sickled red blood cells. Be sure to look for changes in mental status, which can be signs that the blood flow to the brain is being restricted:

- Decreased level of consciousness
- Confusion
- Changes in speech
- Loss of coordination

If sickled cells block blood flow in the brain, it can lead to neurological changes and stroke. Look for changes in mental status, like a decreased level of consciousness, confusion, changes in speech, and loss of coordination.

NURSING INTERVENTIONS

Your top priority for nursing interventions is to maintain adequate oxygenation and blood perfusion to body tissues.

There are 2 underlying causes for most nursing interventions of sickle cell anemia:

- The sickled cells clump together in the blood vessels and block blood flow to the tissues
- The cells are destroyed at a much faster rate than healthy red blood cells

Pain Management as Prescribed

Because all of those sickled red blood cells clumping together and blocking blood flow to body tissues, sickle cell anemia can be very painful. Treating the pain (either acute or chronic) can be done with either IV pain medications or oral pain medications. It may be done with a combination of opioid (morphine, oxycodone) and non-opioid (NSAIDS, acetaminophen) analgesics to help sustain better control.

Patients with sickle cell disease may start to build up a tolerance to the pain

medication regimens, so making sure they are adequately controlled over time is important.

Advocate for your patient to ensure that their pain is properly controlled.

Supplemental Oxygen as Prescribed

Make sure they are getting enough oxygen to their body tissues. Those damaged, sickled red blood cells can't carry oxygen as well as healthy red blood cells, so the body won't be getting as much oxygen as it should.

Supplemental oxygen may be prescribed to help increase the oxygen present for healthy red blood cells to transport, but the sickled cells are still damaged and can't carry enough oxygen, even though we're making oxygen more readily available.

Blood Transfusion as Prescribed

A blood transfusion may be needed and can help to add more healthy red blood cells to carry more oxygen that the sickled cells can't.

Encourage Adequate Hydration

Encourage the patient to stay hydrated. They may be prescribed IV fluids to help increase their fluid volume. This is especially helpful in a vaso-occlusive crisis or a pain crisis, where the sickled cells are blocking blood vessels. The extra fluid helps to dilute the blood and get the blood moving again.

Follow Prescribed Diet Plan

To help the patient's body make more healthy red blood cells, their diet should be:

- High in calories
- High in protein
- High in iron
- High in folic acid

Extend extremities and apply warm compresses as tolerated

Extending the extremities out will help to increase blood flow and decrease swelling. Applying heat to the painful area may also help by dilating the blood vessels and helping get those clumped sickle cells moving again.

Educate on Possible Trigger

Educate your patient on potential triggers of sickle cell crisis:

- Anything that increases oxygen demand since more sickled cells are produced when there is an increased oxygen demand in the body, like
  - Illness
  - Infection
  - Dehydration
  - Stress
  - Extremes in temperature like a fever or exposure to cold temperatures
- Blood loss from surgery or trauma that reduces the amount of healthy red blood cells

Helping your patient understand what can trigger a sickle cell crisis can help them avoid the triggers.

Administer Vaccines as Prescribed

Extending the extremities out will help to increase blood flow and decrease swelling. Applying heat to the painful area may also help by dilating the blood vessels and helping get those clumped sickle cells moving again.

Give Prescribed Medications

The patient may be on antibiotics to help prevent an infection and keep the body healthy so that less sickled cells are made.

Hydroxyurea may be prescribed for a patient with sickle cell disease. This medication helps to increase a certain type of hemoglobin (called fetal hemoglobin or hemoglobin F) and it helps red blood cells keep their normal shape. When giving this medication, watch for infection and bleeding because it can lower the patient's white blood cell and platelet count.

Hydroxyurea can lower the patient's white blood cell and platelet counts. So watch for infection and bleeding.