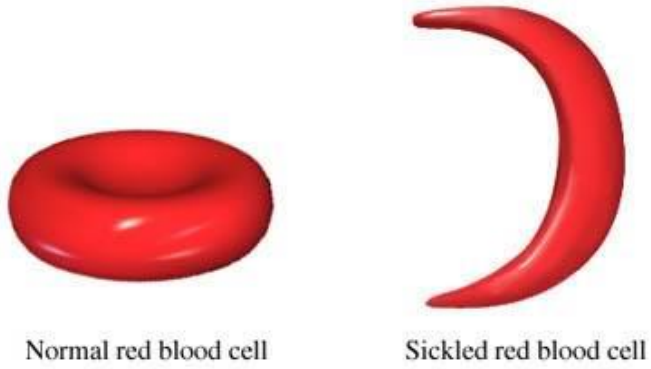
Sickle Cell Disease
Victory Junction Medical Staff and Volunteer Training

Objectives – What's the target?

- Review normal physiology of the blood and clotting systems
- Sickle cell anemia
  - Review basic pathophysiology
  - Discuss common complications and sickle cell emergencies
  - Discuss treatment of sickle cell anemia and complications
  - Discuss considerations for campers
- Bleeding disorders
  - Review basic pathophysiology of the most common bleeding disorders
  - Discuss common complications and emergencies
  - Discuss treatment of bleeding disorders
  - Discuss considerations for campers

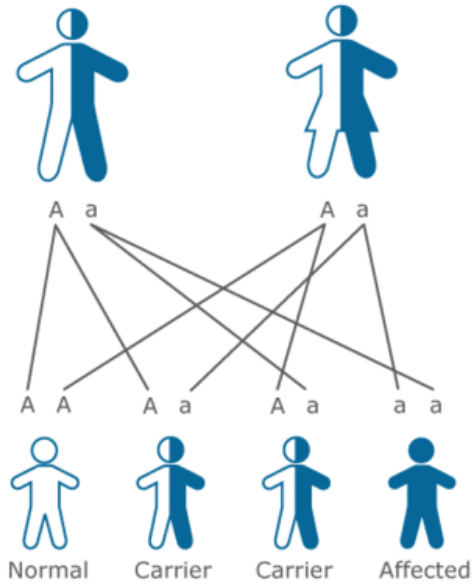
The target is camper safety! Better understanding of the diseases our campers have means better care!

Sickle Cell Disease– What is it?



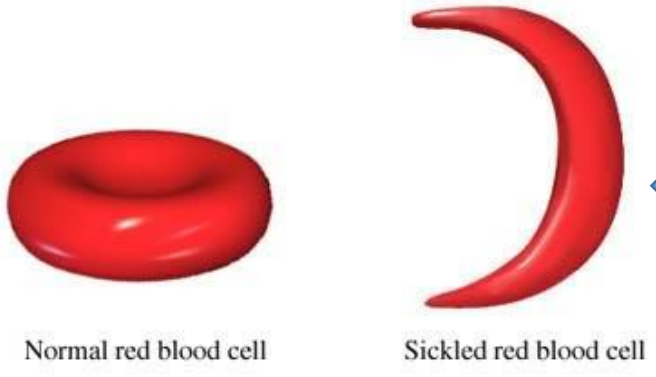
Sickle cell disease (SCD) affects the protein called ***hemoglobin*** (hgb) and a person's red blood cells.

SCD is an inherited, autosomal recessive disease. This means that some people are carriers of the trait and some have the disease. There are also different types of sickle cell disease.



Red Blood Cells

Healthy RBCs are round, smooth, and flexible.



Normal red blood cell

Sickled red blood cell

Sickled RBCs are C-shaped, sticky, and rigid.

*due to a difference in the amino acid chain, the hgb C and S strands create a different shape

Hemoglobin (hgb) is a protein that loosely and reversibly binds with oxygen. Healthy hgb is needed to transport and deliver oxygen to the body.

Normal red blood cells:

- Contain hemoglobin A (normal hgb)
- Carry and deliver oxygen
- Easily flow through blood vessels
- Live for about 120 days

VS

Sickled red blood cells:

- Contain hemoglobin S or C (abnormal hgb)
- Cannot properly carry and deliver oxygen
- Get stuck in small blood vessels
- Live for about 16 days

Sickle Cell Disease – Different Types

SS

HgbSS is the most common and also the most severe form of sickle cell disease, and is called ***sickle cell anemia***. This means a person inherited the sickle "S" gene from both parents.

SC

HgbSC occurs when the "S" gene is inherited from one parent, and a different abnormal hgb "C" is inherited from the other. HgbSC is usually a more mild form of sickle cell disease, but every person is different.

SBetaThal

There are two types of SBetaThal, "0" which is usually a severe form and "+" which is usually mild. "Thal" stands for thalassemia, a different type of anemia inherited from one parent in this type of SCD or it can be a diagnosis without the "S" sickle portion.

Sickle Cell Trait

A person with sickle cell trait (SCT) is a carrier of the disease, but does not have symptoms of the disease.

http://www.cdc.gov/ncbddd/sicklecell/facts.html

Sickle Cell Disease – Demographics

Did you know? All newborns in the U.S. are screened for SCD.

Millions of people worldwide have sickle cell disease.

The CDC estimates that SCD affects 90-100,000 Americans.

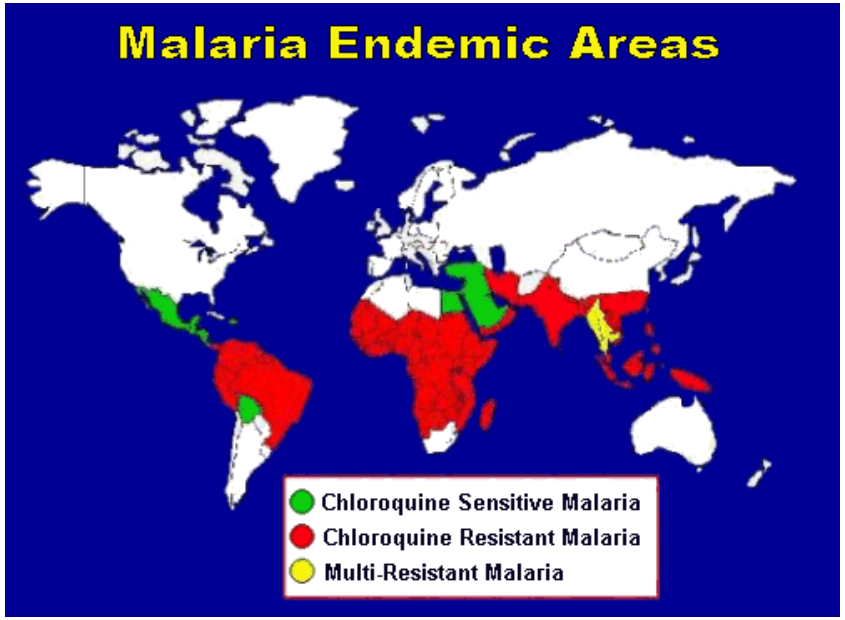
In 2010, approximately 1.5% of babies born in the U.S. tested positive for SCD or SCT (that's 60,000 babies!), including:

73.1 of 1,000 black newborns
6.9 of 1,000 Hispanic newborns

http://www.cdc.gov/mmwr/preview/mmwrhtml/mm6349a3.htm?s_cid=mm6349a3_w

Sickle Cell Disease - Demographics

Sickle cell trait is protective against malaria, so many people living in malaria endemic areas of the world have sickle cell trait.



Knowing that sickle cell disease is inherited, this shows why it has a higher incidence in the African and Hispanic populations.

http://www.cdc.gov/malaria/about/biology/sickle_cell.html

Sickle Cell Disease - Complications

PAIN Stroke Gallstones Jaundice sepsis Vasocclusive crisis organ Priaprism AVN Delayed growth damage Acute Chest Syndrome

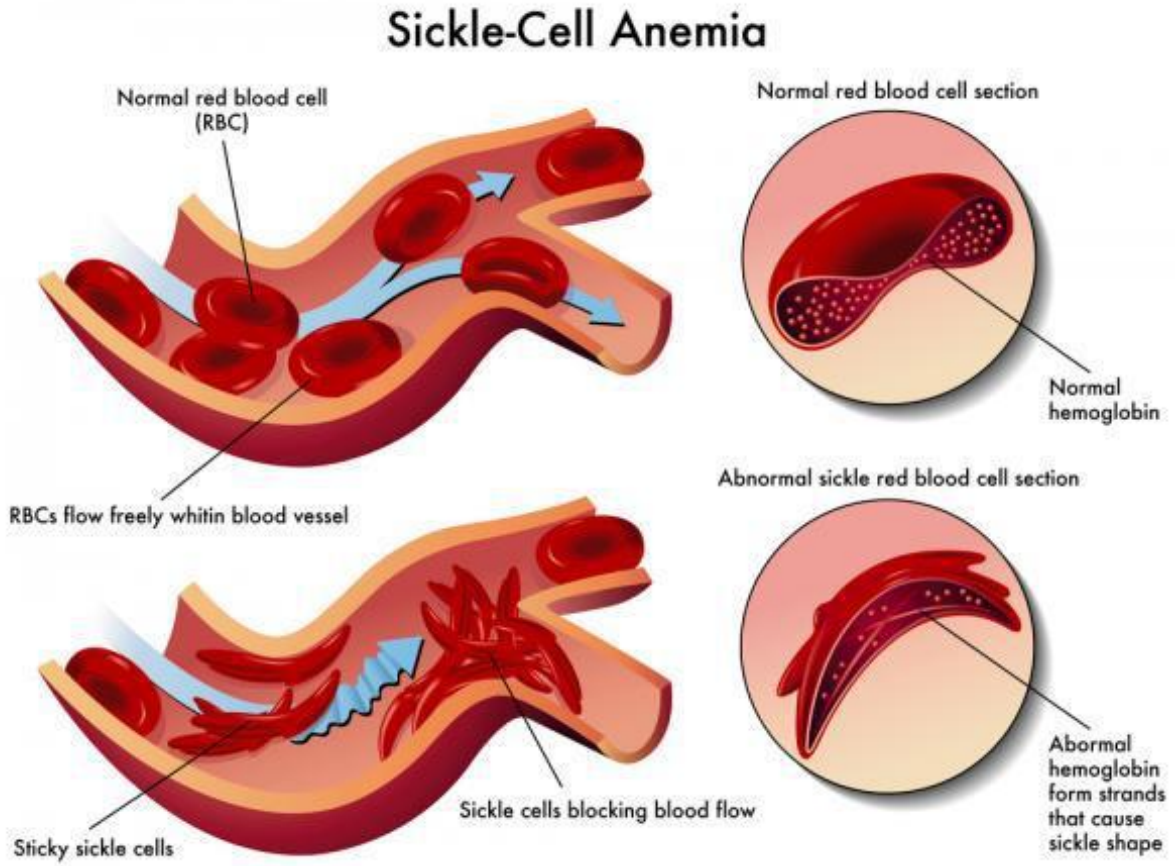
In 1973, the average person with SCD in the U.S. had a lifespan of only **14 years**.

Today, a person with SCD in the U.S. has a life expectancy of **40-60 years**.
The overall life expectancy in the U.S. is **75-80 years**.

http://www.nhlbi.nih.gov/health/health-topics/topics/sca

Sickle Cell Disease – Complications

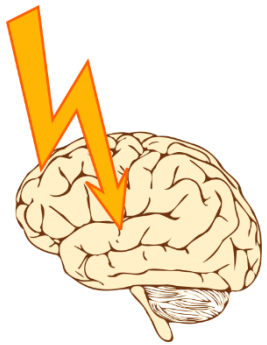
Vaso-occlusive crisis (VOC) – Also called a sickle crisis, this occurs when the sickled cells occlude blood flow, and is responsible for many of the complications of sickle cell disease.



Sickle Cell Disease - Complications

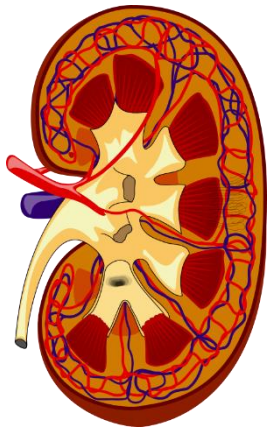
Location, location, location…

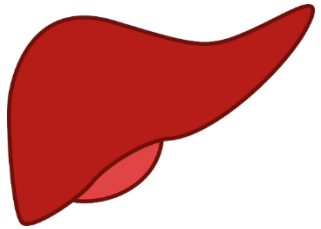
The effect of a vaso-occlusive crisis is determined by the location of the occlusion.



VOC in the Brain = STROKE

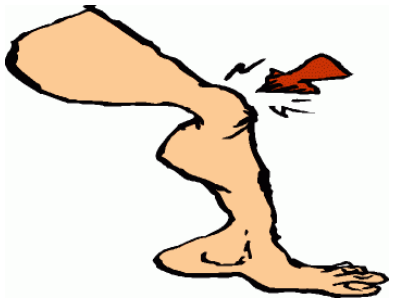
VOC in kidney = organ damage and dysfunction, can lead to renal failure

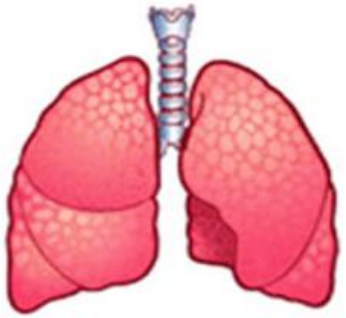




VOC in the spleen = splenic sequestration, organ damage, and functional asplenia (decreased ability to fight certain infections)

VOC in bone, muscle, or joint = PAIN, tissue damage, can lead to avascular necrosis (AVN) of the joint





VOC in lungs = poor oxygenation, Acute Chest Symdrome (ACS)

Sickle Cell Disease – Complications – Stroke

A VOC in the brain blocks blood flow, and oxygen delivery, to the brain tissue and causes a stroke.

Clinical Stroke

- A clinical stroke is one that shows outward symptoms.
  - Differs with location of injury
  - May have altered balance, weakness of one limb or one side of body.
  - May have speech or language difficulties.
- In children, most occur between ages 2 & 9.
- Up to 24% of people with hgbSS may suffer a clinical stroke by age 45.
- Stroke can lead to death.

Silent Stroke

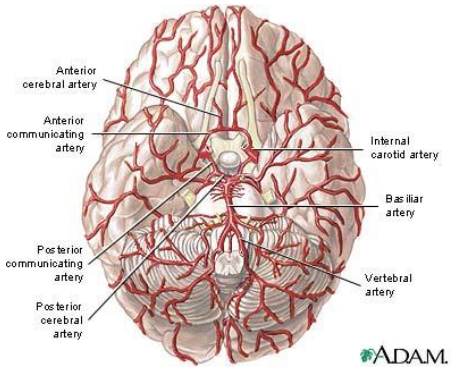
- A silent stroke is brain damage caused by decreased blood flow and oxygen delivery, without outward symptoms.
- This damage leads to cognitive difficulties, and can have an impact on decision making and success in school and work.
- This injury is common, especially in those with hgbSS or hgbSbeta0thal sickle cell disease.

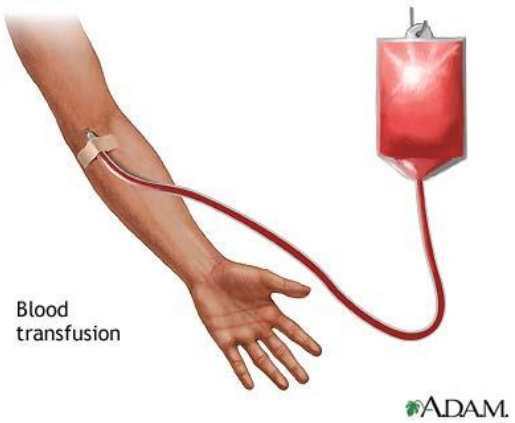
http://www.nhlbi.nih.gov/health/health-topics/topics/sca/signs

Sickle Cell Disease – Complications - Stroke

Stroke risk assessment and prophylaxis has decreased the risk of clinical stroke in children with SCD.

Transcranial Doppler – measures blood flow through blood vessels in the brain to determine a child's risk for stroke





Chronic transfusions can lead to iron overload. Campers may take medicines to help bind the iron and prevent it from depositing in the body and causing organ damage.

Campers on chronic transfusions may have an internal central line, or Port, for easier venous access.

Blood transfusions – infusing healthy red blood cells decreases the percentage of sickled cells circulating and can help prevent stroke in children with high risk. Transfusions may be scheduled on a regular basis (chronic transfusions) or also involve taking out some of the patients sickled blood (exchange transfusions).

http://www.nhlbi.nih.gov/health/health-topics/topics/sca/signs

Sickle Cell Disease – Complications – Organ Damage

Organ damage in sickle cell disease results from lack of oxygen, circulation of abrasive sickled cells, and the by-products of sickled RBCs breaking down more frequently. The kidneys, liver, gallbladder, eyes, and spleen are frequently affected.

Renal Complications

- Sickled cells damage the sensitive and specialized vessels of the kidneys
- The kidneys have trouble concentrating urine.
- Blood and/or protein may be present in the urine
- Later in life, kidney disease and renal failure can occur.

Children with SCD often wet the bed (nocturnal enuresis).

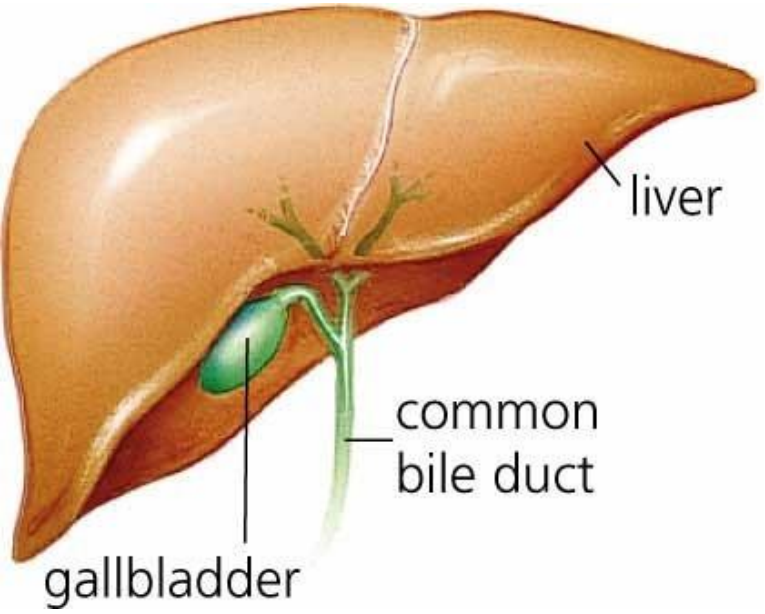
The urine may be dark, due to blood and/or excess bilirubin from RBC breakdown.

Hydration is extremely important for maintaining healthy kidney function in SCD.

Sickle Cell Disease – Complications – Organ Damage

Liver Complications

- In people with iron overload from transfusions, the iron can deposit and damage the liver tissue.
- When sickled cells occlude blood flow and oxygen from reaching liver tissue, it is called "sickle cell intrahepatic cholestasis" and is rare, but sudden and severe.
- Repeated episodes of liver injury can lead to liver failure.



Gallbladder Complications

- When RBCs, including sickled cells, break apart (hemolyze) at the end of the life cycle, the release their hemoglobin, which then breaks down to bilirubin.
- Because sickled cells have a shorter lifespan and break down more frequently, there is more bilirubin than the body can process.
- Excess bilirubin can form stones in the gallbladder, called gallstones, which block the bile ducts.
- Gallstones are common in people with SCD, and many have their gallbladder removed from complications.
  - Symptoms of gallstones include RUQ pain, nausea and vomiting
  - People may be advised to avoid high fat foods.

http://www.nhlbi.nih.gov/health/health-topics/topics/sca/signs

Sickle Cell Disease – Complications – Organ Damage

Eye Complications

- Excess bilirubin may also cause jaundice, often most notable as scleral jaundice (yellowing of the eyes).
- Retinal damage from can lead to blindness.
  - Vascular damage from abrasive sickled cells
  - Overgrowth of blood vessels to compensate can also damage the retina

Priaprism

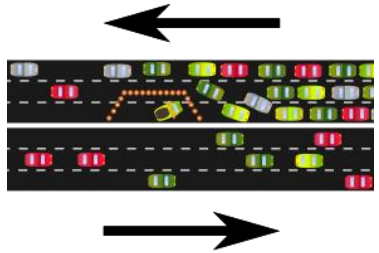
- In males with SCD, a VOC in the vessels of the penis can lead to prolonged, unwanted, extremely painful erection
- An erection lasting > 4 hours must be treated as an emergency
- Permanent damage to the penis can occur
- Priaprism is often treated with Sudafed and heat to increase vasodilation, but can require surgical intervention

http://www.nhlbi.nih.gov/health/health-topics/topics/sca/signs

Sickle Cell Disease – Complications – Organ Damage

Spleen Complications

- The spleen plays an important role in the immune system by filtering blood cells and bacteria. The spleen is often damaged in SCD.
- ***Splenic sequestration*** occurs when RBCs get "stuck' in the spleen, causing it to enlarge quickly and the circulating RBC/hgb levels to decrease rapidly.
  - This can quickly become an emergency in children.
  - Children with severe or repeated splenic sequestration may have the spleen removed.



In splenic sequestration, red blood cells get "backed up" like a traffic jam in the spleen.

Fever in a camper with SCD is an emergency!

- ***Functional asplenia*** occurs when the spleen is present, but does not function.
  - This is common in children and adults with sickle cell disease.
  - Children are often treated with penicillin prophylaxis.
  - Infection and fever in a person with SCD can be an emergency due to their decreased ability to fight serious bacterial infections such as:
    - Pneumococcus, Haemophilus influenza type B, Meningococcus, Salmonella, Staphylococcus, Chlamydia, *Mycoplasma pneumoniae*

http://www.nhlbi.nih.gov/health/health-topics/topics/sca/signs

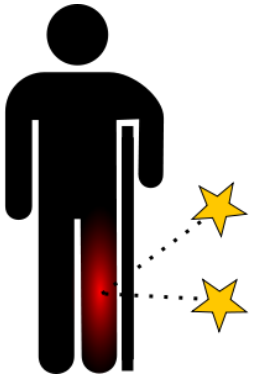
Sickle Cell Disease – Complications - Pain

Pain is the most common and widely recognized complication of SCD.

People with SCD can have acute pain, chronic pain, or both.

Pain associated with VOC in the bones, muscles, and joints is a result of tissue ischemia and inflammation around the site of vaso-occlusion.

- Sites of VOC related pain are often warm to the touch, swollen, and have various alleviating and aggravating factors.



Pain can occur anywhere – legs, hips, and back are common sites.

Sickle Cell Disease – Complications - Pain

Assessing pain in children with a chronic illness can be challenging, but
Even if he wants to play a game…
Even if he laughs at a joke…
Even if he looks fine…

Believe the camper when he says he has pain!

Some people with SCD have chronic pain, and their baseline may be a pain level other than 0. The goal for pain relief may not always be a "0."

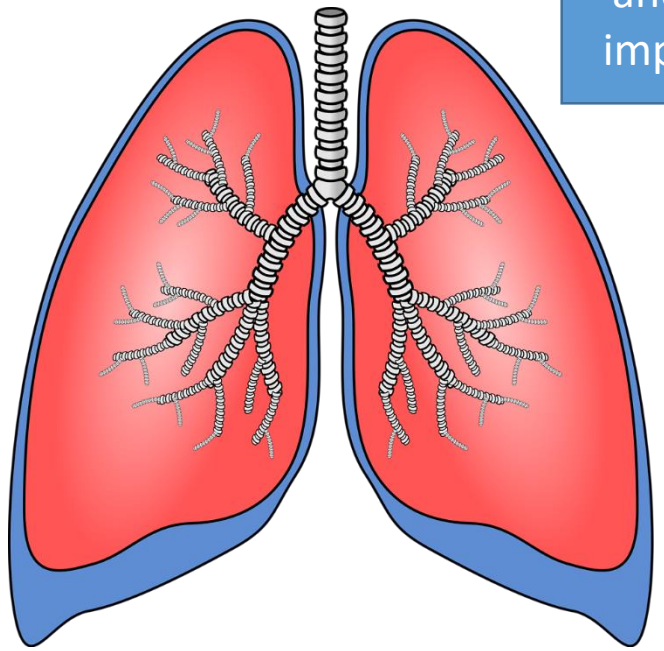
Pain assessment includes:

- Onset
- Intensity
- Character (burning, stinging, sharp, etc.)
- Aggravating and alleviating factors
- Re-assessment with interventions

Sickle Cell Disease – Complications – Acute Chest Syndrome

Acute Chest Syndrome (ACS) is the most common pulmonary complication of SCD, and the leading cause of death in people with SCD.

Acute Chest Syndrome is defined by a ***new infiltrate on a chest x-ray*** with ***fever*** and/or ***respiratory symptoms*** (most often cough).



Early intervention and treatment is important in ACS.

ACS is classified as mild, moderate or severe

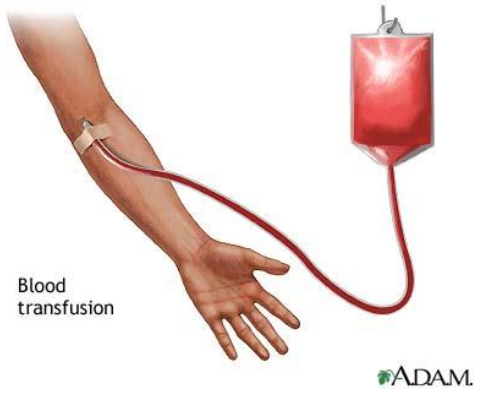
Adults are more likely to have severe ACS and higher mortality rates from ACS

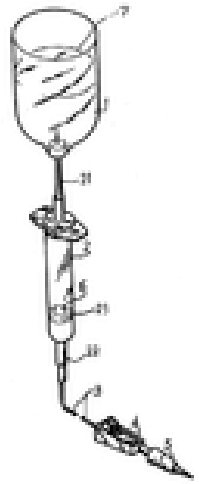
http://www.uptodate.com/contents/acute-chest-syndrome-in-adults-with-sickle-cell-disease?source=see_link

Sickle Cell Disease – Complications – Acute Chest Syndrome

About half the cases of ACS develop during hospitalization for VOC/pain, surgery, or asthma. These are risk factors for developing ACS.

Treatment of ACS may include respiratory support (beginning with oxygen and incentive spirometry and progressing to mechanical ventilation), administration of IV fluids and antibiotics (if applicable), or blood transfusions.





In children, the cause of ACS is often a viral respiratory infection.

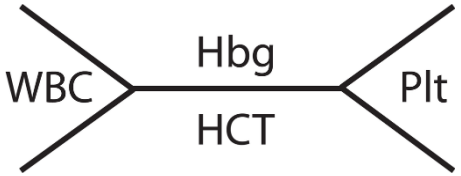
In adults, the cause of ACS is more often bone marrow or fat emboli.

Sickle Cell Disease – Complications - Anemia

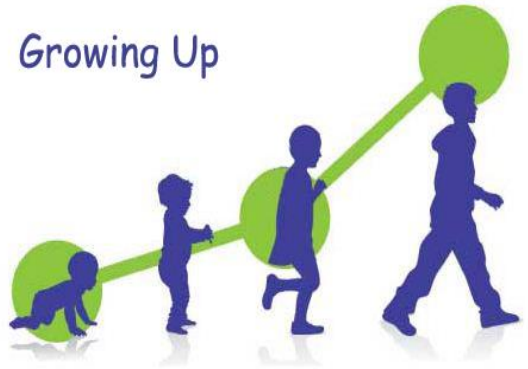
Anemia is a well-known symptom of SCD, especially HgbSS which is called “sickle cell anemia.”

Symptoms of chronic anemia include

- Fatigue
- Headaches
- Pallor
- Chronic hypoxia/hypoxemia
- Delayed growth and development



Baseline hgb levels in a person with SCD can be as low as 6 g/dl, or at a normal level of 14-16 g/dl.



Sickle Cell Disease - Treatments

People with sickle cell disease take medications to control symptoms and prevent complications. Some of the most common include:

Folic Acid – promotes RBC production. Most children will take a 1mg tab once a day.

Penicillin ("PenVK")– prophylaxis against bacterial infections due to immune (spleen) dysfunction
-Usually taken as "PenVK" or penicillin with potassium, once or twice a day.
*Guidelines vary on the recommended age to discontinue this prophylaxis, and vary depending on whether the child's spleen has been removed.

Deferasirox ("Exjade") – chelation treatment for iron overload due to blood transfusions.
Taken as tablets dissolved into liquid (often orange juice), it dissolves faster in a little bit of warm water, then mixed with juice. Do not mix with a metal spoon.
*this tastes horrible and many kids hate taking it

The only curative treatment for SCD is a bone marrow transplant. While there is a great benefit, this treatment also has many very serious risks.

Sickle Cell Disease - Treatments

Hydroxyurea (also "Hydrea" or "Droxia") – a chemotherapy medication that promotes production of fetal hemoglobin. Fetal hgb doesn't sickle, and use of hydroxyurea has become much more common recently with positive results in reducing VOC!

Hydroxyurea is a chemotherapeutic agent. The pill/liquid bottle will be labeled as a biohazard.

When giving this medication at camp:

- Verify the 5 rights of med administration!
- Wear gloves if directly handling the medication
- Wash hands immediately with soap and water if your skin came into contact with the medication. (Campers will touch their medication, hand washing is encouraged for everyone.)
- Dispose of liquid medication syringes in the labeled YELLOW biohazard bin.
- If giving the liquid suspension, ensure it doesn't spill – if it does, clean the area immediately with CHG wipes.

Some campers will always take hydroxyurea on an empty stomach, some with food. Keep the routine at camp.

It is often taken at night due to the side effect of nausea. (typically mild)

Chemo certification is not required to administer hydroxyurea at camp. If you have concerns, please ask the full time staff.

Sickle Cell Disease at Camp

How to keep your campers with sickle cell disease safe and healthy:

HYDRATE!!!!
Hydration is key for preventing VOC.

Stay warm and dry! Abrupt change in temperature and cold can trigger VOC.

Treat pain early. Hydrate, rest and apply heat at the first signs of VOC pain.

Get campers into the warming hut to dry ***immediately*** after swimming in the heated pool.

No ice packs! On rainy days, ensure campers stay dry or change out of wet clothes.

Sickle Cell Disease at Camp – Pain Management

We do our very best to keep campers healthy, but it is likely that you will have a camper with sickle cell disease experience pain during the session. This is an example of how we manage VOC pain in the Body Shop or at camp (subject to change).

- Believe the camper when he says he has pain.
- Assess the camper. Assess for fever, change in respiratory status, and evaluate the site of pain.
  - Pain can be an initial symptom of an illness, always assess for symptoms of infection.
  - For joint/muscle pain, attempt to assess whether it is VOC related or strained/other.
  - It is ok to ask the camper if the pain "feels like sickle cell pain" and what he would usually do
- Initially treat with rest, oral hydration, and heat applied to the site of pain.
  - Assess the camper's hydration status – When did he last void? How much water has he had to drink today?
  - No ice packs.
- First line medication is almost always ibuprofen or another NSAID. This reduces pain and promotes vasodilation.
- If pain is not improved, have the provider on call assess the camper and treat according to "pain plan." (example below)

Mild pain (0-3)
- Hydrate
- Ibuprofen 400mg PO q8hrs
- Heat packs

Moderate pain (4-7)
- Continue ibuprofen 400mg PO q8hrs
- Oxycodone 5mg PO Q4hrs PRN
- IV hydration

Severe pain (7-10)
- Change ibuprofen to ketorolac (Toradol) 20mg IV q6hrs
- Morphine 2mg IV q3hrs PRN
- IV hydration

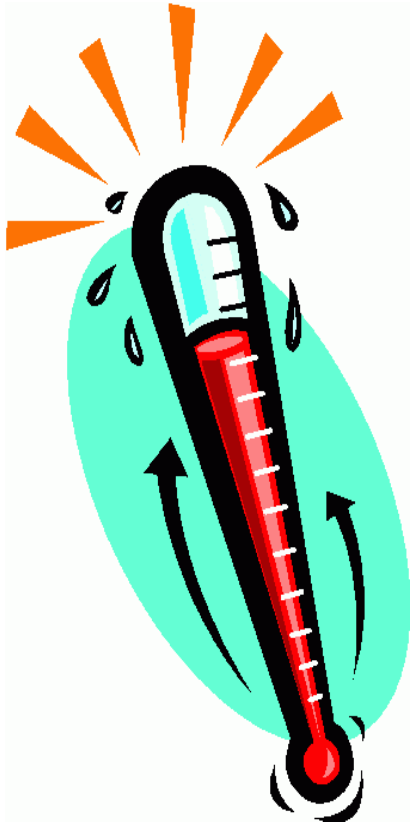
**Note on Abdominal pain

Constipation is common! (at camp and in kids with SCD)

Sickle Cell Disease at Camp – Fever Management

We do our very best to keep campers healthy, but it is possible that you will have a camper with sickle cell disease develop a fever during the session.

- Assess for fever when a camper with SCD is feeling unwell, or has a headache/nausea/pain etc.
- Fever in a child with SCD is typically any temperature >38.5 C or >101.3 F
- Assess the camper (VS, respiratory status, brief head to toe), and **notify the provider on call.**
- Anticipate drawing labs (blood culture, CBC at minimum).
- Anticipate the provider's orders for IV antibiotics and fluids if needed.



Remember: Ask for help! You will never be asked or expected to manage this type of situation alone.

Sickle Cell Disease - Emergencies

Conditions requiring immediate medical attention include:

- Signs of stroke, any neuro changes
- Respiratory distress
- Fever
- Pain that does not improve with initial treatments
- Priaprism lasting > 4hrs

During medical check-in, ask your camper

- Have you had pain recently?
- When you have pain, where is it most often?
- How do you treat pain at home? What works for you?

Helpful things to know about your campers with sickle cell disease:

- Baseline SpO2 levels
- Baseline Hgb level
- History of complications/hospitalizations
- Usual site of pain/VOC
- Individual pain plan and coping

Sickle Cell Disease - Emergencies

Conditions requiring immediate medical attention include:

- Signs of stroke, any neuro changes
- Respiratory distress
- Fever
- Pain that does not improve with initial treatments
- Priaprism lasting > 4hrs

Helpful things to know about your campers with sickle cell disease:

- Baseline SpO2 levels
- Baseline hgb level
- History of complications/hospitalizations
- Usual site of pain/VOC
- Individual pain plan and coping

Pain is likely to be the most common concern at camp. Anticipate managing with:

- Hydration
- Heat applied locally – this provides comfort and encourages vasodilation
- Anti-inflammatory medications if no contraindications
- Distraction, relaxation, music, and support from counselors
- Oral or IV narcotics if ordered

- Remember that a child with SCD in VOC is at higher risk for developing ACS, assess the child's respiratory status and vital signs periodically.

Thank you!