



Sickle Cell Disease

OBJECTIVE

The objective of this Clinical Practice Guideline (CPG) is to provide evidence-based practice recommendations for the treatment of sickle cell disease. The CPG discusses goals and key treatment plan components for members with sickle cell disease, treating the disease while pregnant, as well as behavioral health implications. In addition, the CPG outlines the organizations that WellCare aligns with regarding sickle cell disease and Measureable Health Outcomes.

OVERVIEW

Sickle cell disease is a genetic disorder that affects the body's red blood cells. In this disease, defective hemoglobin (a substance that carries oxygen in the blood) causes the red blood cells to change shape (into a sickle) when oxygen is released to tissues. Normal red blood cells are round and are able to move through small blood vessels in the body to deliver oxygen. In sickle cell disease, a chemical change in hemoglobin causes the substance to form long rods in the red blood cell as the hemoglobin releases oxygen. These rigid rods change the shape of the red blood cell into a sickle shape, hence the name of the disease.¹ Acute painful sickle cell episodes (or painful crises) are caused by blockage of the small blood vessels. The red blood cells in people with sickle cell disease behave differently under a variety of conditions, including dehydration, low oxygen levels and elevated temperature. Changes in any of these conditions may cause the cells to block small blood vessels and cause tissue infarction. Repeated episodes may result in organ damage. Episodes occur unpredictably, often without clear precipitating factors. The frequency varies from less than one episode a year to severe pain at least once a week. Pain can fluctuate in both intensity and duration, and may be excruciating. The majority of painful episodes are managed at home, with patients usually seeking hospital care only if the pain is uncontrolled or they have no access to analgesia. Those requiring admission may remain hospitalized for several days. The primary goal to managing an acute painful sickle cell episode is to achieve effective pain control promptly and safely.² Individuals with sickle cell disease may have hemoglobin "S", SC, S-beta thalassemia and others genetically distinct conditions, whereas others without this disease have hemoglobin "A". Sickle cell disease is found most often in African-Americans and Africans. However, other ethnic groups also can have sickle cell disease.¹

Trait. Sickle cell is a benign carrier condition, usually with none of the symptoms of sickle cell anemia. It is generally only a laboratory diagnosis in which one sickle cell gene has been inherited along with a normal gene.¹

Population. There are approximately 2.5 million people in the United States and 300 million in the world who have sickle cell trait. Everyone with sickle cell trait needs genetic counseling (not necessarily from a geneticist), regarding the risk of having a child with sickle cell disease.¹

Presence, Geographic Coverage and Prevalence. The presence of sickle cell trait appears to be protective against severe falciparum malaria, explaining the persistence of this gene in the world population. The gene for sickle cell anemia is widespread throughout the world, being present in Africa, Mediterranean countries (especially Greece), the Middle East, and parts of India. The prevalence of sickle cell trait is approximately 8 to 10 percent in African Americans and as high as 25 to 30 percent in certain areas of western Africa. The Hispanic population is also at risk for sickle cell disease. At least 1 in 180 Hispanic births have sickle cell trait.¹

Hierarchy of Support

GUIDELINE HIERARCHY

CPGs are updated annually or as necessary due to updates made to guidelines or recommendations by the National Heart Lung and Blood Institute (NHLBI), National Institute for Health and Care Excellence (NICE), and the American Academy of Pediatrics (AAP). When there are differing opinions noted by national organizations, WellCare will default to the member's benefit structure as deemed by state contracts and Medicaid / Medicare regulations. If there is no specific language pertaining to sickle cell disease, WellCare will default (in order) to the following:

- National Committee for Quality Assurance (NCQA);
- United States Preventive Services Task Force (USPSTF), National Quality Strategy (NQS), Agency for Healthcare Research and Quality (AHRQ);
- Specialty associations, colleges, societies, etc. (e.g., American Academy of Family Physicians, American Congress of Obstetricians and Gynecologists, American Cancer Society, etc.).

Links to websites within the CPGs are provided for the convenience of Providers. Listings do not imply endorsement by WellCare of the information contained on these websites. NOTE: All links are current and accessible at the time of MPC approval.

WellCare aligns with the NHLBI, NICE, and AAP on the topic of sickle cell disease. Highlights from their respective publications are noted below.

NATIONAL HEART, LUNG, AND BLOOD INSTITUTE (NHLBI)

The National Heart Lung and Blood Institute (NHLBI) published guidelines in 2014 that align with scientific evidence in the following areas:³

- Penicillin prophylaxis prevents pneumococcal sepsis in children;
- Pneumococcal vaccine prevents pneumococcal infection in children;
- In surgical settings, simple transfusions to increase hemoglobin (Hb) levels to 10 g/dL are as good as or safer than aggressive transfusions to reduce sickle hemoglobin (Hb S) levels to below 30 percent;
- Transfusions to maintain a hematocrit of more than 36 percent do not reduce complications of pregnancy;
- Transfusions to reduce Hb S levels to below 30 percent prevent strokes in children with high central nervous system blood flow; and
- Hydroxyurea decreases crises in patients with severe sickle cell disease.

For recommendations related to reproductive counseling and care during pregnancy for women with the sickle cell trait or the disease, refer to NHLBI's *Evidence-Based Management of Sickle Cell Disease - Expert Panel Report, 2014* at: <http://www.nhlbi.nih.gov/sites/www.nhlbi.nih.gov/files/sickle-cell-disease-report.pdf>.

Additional topics covered by the NHLBI guideline include health maintenance, management of acute and chronic conditions, use of hydroxyurea, and blood transfusions in the management of SCD. The full NHLBI guidelines are available at https://www.aap.org/en-us/Documents/soho_clinical_topic_sickle_cell.pdf.

NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE (NICE)

The National Institute for Health and Care Excellence (NICE) published recommendations for managing acute painful episodes in the hospital. Recommendations focus on:²

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| • Individualized assessment at presentation | • Management of underlying pathology |
| • Primary analgesia | • Non-pharmacological interventions |
| • Reassessment and ongoing management | • Settings and training |
| • Possible acute complications | • Discharge information |

For the full list of recommendations, please visit the NICE website <https://www.nice.org.uk/guidance/cg143/chapter/1-recommendations>.

AMERICAN ACADEMY OF PEDIATRICS (AAP)

In 2011, the American Academy of Pediatrics reaffirmed their 2002 guideline. The guidelines provide pediatricians in primary care and subspecialty practice with an overview of the genetics, diagnosis, clinical manifestations, and treatment of sickle cell disease. The provision of comprehensive care is a time-intensive endeavor that includes ongoing patient and family education, periodic comprehensive evaluations and other disease-specific health maintenance services, psychosocial care, and genetic counseling. Timely and appropriate treatment of acute illness is critical, because life-threatening complications develop rapidly. It is essential that every child with SCD receive comprehensive care that is coordinated through a medical home with appropriate expertise.⁵ Health maintenance for pediatric members should be comprehensive, ensuring that providers discuss the following with the member and/or caregiver: prophylactic medications, immunizations, comprehensive medical evaluation and psychosocial care. Further, the AAP recommends that health supervision should consist of family education, health maintenance, acute illness and psychosocial care. The AAP's report can be found at <http://pediatrics.aappublications.org/content/109/3/526.full>.⁴

Evidence Based Practice**MEASUREMENT OF COMPLIANCE**

WellCare is committed to adhering to the measures and standards published by the Centers for Medicare and Medicaid Services (CMS) and the National Committee for Quality Assurance (NCQA). Please reference WellCare's Clinical Policy Guiding Document titled *Measures of Compliance*.

NOTE: To access Clinical Policy Guiding Documents visit www.wellcare.com – select the Provider tab, then “Tools” and “Clinical Guidelines”.

Care Management

The goals for Care Management is to support the member and caregiver's ability to self-manage sickle cell disease, minimize risks factors, and remove barriers preventing the member from achieving those goals. Members and their caregiver(s) should be educated on the items below related to the primary symptoms of sickle cell disease.

Members (or their caregiver) should call a Provider right away to report increased signs of anemia:^{7,8}

- Tiredness
- Irritability
- Dizziness and lightheadedness
- Fast heart rate
- Difficulty breathing
- Pale skin color
- Jaundice (yellowish tint to skin, whites of eyes)
- Delayed healing
- Painful swelling of fingers and toes

Parent(s)/caregiver(s) should talk to the child's physician if the child doesn't seem to be growing or developing at the same rate as other children of the same age.^{7,8}

Members should seek immediate medical care for symptoms of a sickle cell crisis:^{7,8}

- Fever
- Shortness of breath
- Chest pain
- Rapid breathing
- Cough
- Weakness of an arm, leg or one side of the body
- Trouble speaking, walking or understanding
- Loss of balance
- Severe headache
- Pain in the eye or sudden visual changes
- Blood in the urine
- Changes in how often and amount of urination

Integrated care management of sickle cell involves:

- Coaching related to stress management skills
- Coaching related to pain management skills
- Coaching related to coping skills
- Strategies for pain management, including non-pharmacological
- For pediatric members, monitoring of pediatric developmental milestones
- Vaccinating against influenza, pneumonia and meningitis, and consider penicillin therapy especially in ages 0-5
- Including as part of routine assessments: oxygen saturation, eye exams, and Transcranial Doppler (TCD)

Ultrasound Screening, if appropriate.

For additional information related to the signs and symptoms of sickle cell disease, please see the Addendum.

MEASURABLE HEALTH OUTCOMES

Targeted Health Outcomes (Extended Program Goals) result from successful member self-management (see Case Management Objectives).

1. The member is able to maintain or improve pain control over a specific period of time after the start of Case Management engagement. Compare pain assessment responses, initial to subsequent assessments.
2. The member experiences less frequent instances of complications (anemia, acute chest syndrome, pain crisis, splenic sequestration, vascular blockages) requiring acute medical attention over a specific period of time. Compare frequency of acute utilization for these complications pre- and post-engagement. Monitor for ED and inpatient authorization/utilization related to the primary diagnosis of sickle cell or related complications. In absence of ED and inpatient utilization, authorizations and claims data, or to otherwise demonstrate the member has not required acute medical intervention, CM may use Provider and/or Member narrative.
3. The member experiences fewer instances of infection requiring acute medical attention over a specific period of time. Compare frequency of acute utilization for infection pre- and post-engagement. Monitor for ED and inpatient authorization/utilization related to a primary diagnosis of infection. In absence of ED and inpatient utilization, authorizations and claims data, or to otherwise demonstrate the member has not required acute medical intervention for infection, CM may use Provider and/or Member narrative.

CASE MANAGEMENT GOALS

Case Goals should target specific care gaps and/or adherence issues, and measure the member's progress towards self-management and adherence which will lead to the targeted health outcomes above. Examples:

1. Member describes fluid intake that meet physician-prescribed (and / or nationally recognized care standard) hydration requirements over the last 30 days and describes risks associated with inadequate fluid intake.
2. Member describes the use of infection prevention measures (such as handwashing and food cleansing) over the last 30 days and has received annual flu vaccine and pneumococcal vaccine if appropriate for age and risk factors.
3. Member is able to avoid cold exposure, high altitudes, and other situations that could decrease circulation in extremities and cause low blood oxygen levels, over the last 30 days.
4. Specific for Members requiring hospitalization: The Member participates in provider follow-up visit within 7 days of hospital discharge.

CASE MANAGEMENT OBJECTIVES

Case Management Objectives should focus on improving the member's self-management skills including:

- Early identification of warning symptoms of complications requiring a call to the physician
- Early identification of crisis situations requiring a call to emergency services
- Recognizing signs of infection to report to physician
- Ways to prevent infection, like frequent handwashing, food safety, and risk/age-appropriate vaccinations
- Pain management techniques, including pharmacological and non-pharmacological
- Managing stress
- Drinking enough fluid each day (4-6 liters for adults, or other amount prescribed by physician or pediatrician)
- Dressing appropriately for the weather and trying not to get too hot or too cold
- Avoiding places or situations that expose you to high altitudes (example: flying, mountain climbing)
- Avoiding places or situations that cause low blood oxygen levels (example: exercising extremely hard)
- Avoiding second-hand smoke
- Taking medications as prescribed
- Adhering to regular provider visit(s) as scheduled (see *Pediatric Preventive Health: HS-1019*)
- Maintaining a healthy body weight

MEDICAL AND BEHAVIORAL HEALTH INTEGRATION

Children with sickle cell disease are 2 to 3 times more likely than their healthy peers to have mental health problems, a feature that is consistent with children with any chronic illness. In addition to an increase in mental health issues for the sickle cell child, nearly half of their mothers also reported significant symptoms of stress.¹

Hospitalization is an especially hard time for children with sickle cell disease and their parents. The school-aged child with sickle cell disease, on average, will miss 29 days of school per year due to hospitalizations and home recovery. This creates challenges for the child, parents, social workers and educators. Treatment planning should therefore also include education issues.¹

Financial support to these families frequently becomes necessary because of the economic hardship that often accompanies hospitalization. Because of medical leave related to the child's school absence or hospitalization, many families experience employment-related issues.¹

Denial. Children with sickle cell anemia are well most of the time, but certain complications can occur which are very serious and sometimes fatal (cause death). It may be hard to believe that a healthy looking child has a life threatening disease, but this is true when a child has sickle cell anemia. Parents may initially show denial of their child's condition, but as episodes recur, the denial will break down.¹

Anxiety, Depression and Other Symptoms. The most frequent psychological problems encountered include increased anxiety, depression, social withdrawal, aggression, poor relationships and poor school performance. High levels of parental anxiety, overprotection, excessive feelings of responsibility and guilt. Children with sickle cell disease are also at high risk for neurological complications including strokes and silent infarcts of the brain. When the brain is impacted there are usually impairments in verbal, performance, and IQ measures. These cognitive deficits may further affect the child's ability to manage pain, stress, and following a treatment plan.¹

Addiction and Substance Abuse in Patients with Sickle Cell Disease. Sickle cell does not predispose to or provide absolute protection from drug addiction. Treatment of patients with drug addiction and a sickle cell syndrome poses a number of very difficult management problems.¹

1. Drug addiction compromises the ability to diagnose and treat complications in the disease.
2. Pain episodes are less responsive to therapy in drug tolerant patients.
3. Drug seeking behavior may mask symptoms of serious complications.
4. Regular use of opiates may make the health of sickle syndrome patients worse.

Motivational Interviewing should be incorporated in assessing the member's readiness to address substance use behaviors. Sickle cell syndromes complicate the treatment of narcotic addiction. It is not realistic to expect these patients to achieve the goal of many drug treatment programs which is total narcotic free status. Pain episodes may require administration of opiate analgesics on an infrequent basis for a short period of time, even in the setting of known addiction. Some patients may have complications that cause chronic pain which will be unmanageable with non-opiates once the individual has become use to the relief obtained by regular opiate use for prolonged period of time.¹

This treatment plan forms a basis for a formal contract which defines the treatment plan, health care givers and patient responsibilities, treatment alternatives, and consequences of non-compliance. Although it is acknowledged that many patients in this addicted group do not want specific treatment for addiction, a positive, constructive approach is required to provide adequate comprehensive medical management of patients with sickle syndromes and iatrogenic drug addiction. Addiction in the sickle cell patient must be addressed with a positive and constructive approach. Principles established in the treatment of addiction are incorporated into an individual treatment plan that takes the individual's strengths, problems, and needs into account.¹

OTHER CONSIDERATIONS

Complications of Sickle Cell Disease. Symptoms of SCD vary and typically begin during the first year of life, approximately around 5 months of age. Treatment options are different for each person depending on the symptoms. The only cure for SCD is a bone marrow or stem cell transplant. Common complications include:^{1,3,4}

- Hand-Foot Syndrome
- Pain "Episode" or "Crisis"
- Anemia
- Infection
- Acute Chest Syndrome
- Splenic Sequestration
- Vision Loss
- Leg Ulcers
- Stroke
- Deep Vein Thrombosis (DVT) and Pulmonary Embolism (PE)

Other possible complications of SCD include:⁹

- Damage to body organs (like the liver, heart, or kidneys), tissues, or bones due to a lack of blood flow
- Malnutrition / growth retardation among adolescents can cause a delayed onset of puberty; infertility in males
- Gallstones
- Painful erection of the penis (priapism) can last ≤ 2 hours or ≥ 4 hours and may lead to impotence

MEMBER EDUCATIONAL RESOURCES

WellCare contracts with Krames/StayWell for Member educational materials utilized by Case Managers. Items are available to review with Members to address knowledge gaps and verbally educate Members on the topics below:

- Sickle Cell Disease
- Sickle Cell Disease in Children
- Sickle Cell Disease and Pregnancy
- Home Care for Children with Sickle Cell Disease
- Treating Sickle Cell Anemia in Children
- When Your Child Has Sickle Cell Anemia
- What Are Red Blood Cells?
- Hemoglobin S
- Autosomal Recessive Inheritance

These materials are in the approval process and will be available for member educational mailing in the future. Providers may wish to research the titles above related to sickle cell disease that Case Managers utilize with Members. Additional information for Members and their caregiver(s) can be found in the Addendum.

Related WellCare Guidelines

In addition to the information contained in this document, please reference the following CPG: *Pediatric Preventive Health: HS-1019*.

NOTE: Clinical Policies can be accessed by going to www.wellcare.com – select the Provider tab, then “Tools” and “Clinical Guidelines”.

References

1. WellCare. Internal Care Management Training. 2015.
2. Sickle cell disease: managing acute painful episodes in hospital (clinical guideline CG143). National Institute for Health and Care (NICE) Excellence Web site. <https://www.nice.org.uk/guidance/cg143/chapter/1-recommendations>. Published June 2012. Accessed November 11, 2016.
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4. American Academy of Pediatrics. Health supervision for children with sickle cell disease [reaffirmed 2011]. *Pediatrics*, 109(3), 526-535. Retrieved from <http://pediatrics.aappublications.org/content/109/3/526.full>.
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7. What are the signs and symptoms of sickle cell disease? National Health, Lung, and Blood Institute Web site. <http://www.nhlbi.nih.gov/health/health-topics/topics/sca/signs>. Published August 2, 2016. Accessed November 11, 2016.
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9. Sickle cell disease: complications and treatments. Centers for Disease Control and Prevention Web site. <http://www.cdc.gov/ncbddd/sicklecell/treatments.html>. Published August 31, 2016. Accessed November 14, 2016.
10. Living well with sickle cell disease. Centers for Disease Control and Prevention Web site. <http://www.cdc.gov/ncbddd/sicklecell/healthyliving-living-well.html>. Published August 31, 2016. Accessed November 14, 2016.
11. Emergency guide: when to see the doctor. Centers for Disease Control and Prevention Web site. <http://www.cdc.gov/ncbddd/sicklecell/healthyliving-emer-guide.html>. Published August 31, 2016. Accessed November 14, 2016.
12. Five tips to help prevent infections. Centers for Disease Control and Prevention Web site. <http://www.cdc.gov/ncbddd/sicklecell/healthyliving->

[prevent-infection.html](#). Published August 31, 2016. Accessed November 14, 2016.

13. Facts about sickle cell disease. Centers for Disease Control and Prevention Web site. <http://www.cdc.gov/ncbddd/sicklecell/facts.html>. Published August 31, 2016. Accessed November 14, 2016.

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*Easy Choice Health Plan – Harmony Health Plan of Illinois – Missouri Care – Ohana Health Plan, a plan offered by WellCare Health Insurance of Arizona – Staywell of Florida
WellCare (Arkansas, Connecticut, Florida, Georgia, Illinois, Kentucky, Louisiana, Mississippi, Nebraska, New Jersey, New York, South Carolina, Tennessee, Texas) – WellCare Prescription Insurance*

Medical Policy Committee Approval History

Date	History and Revisions by the Medical Policy Committee
10/10/2017	<ul style="list-style-type: none"> • Approved by MPC. No changes.
12/8/2016	<ul style="list-style-type: none"> • Approved by MPC. Enhanced Care Management and Measures of Compliance sections. Revised with CM, DM, QI, UM, BH and the Chief Medical Directors.
2/5/2015	<ul style="list-style-type: none"> • Approved by MPC. Inclusion of <i>Care Management</i> items.
6/17/2014	<ul style="list-style-type: none"> • Approved by MPC. New.

Addendum

LIVING WITH SCD

Providers can encourage parents and caregivers raising a child with SCD to keep in mind the following:¹

- Parents should discourage a "sick person" identity and instead, encourage regular lifestyle.
- No special diet is needed. Encourage a healthy balanced diet.
- Discipline should be administered the same as any other child.
- The importance of school work and a good education should be stressed.
- A regiment of regular follow up with the patient's medical team should be scheduled.
- It is important for the parent and child to know that there can be a delay as long as three years in reaching full adult development. The adolescent with sickle cell anemia may still look and feel like a child while his or her friends are developing adult characteristics. Reassurance that they too will grow up and mature will be necessary to relieve anxious feelings.

Sickle cell disease is a complex hereditary disease. Quality medical care takes a comprehensive team of doctors, nurses and health care professionals working together. Your doctor should have a thorough knowledge of sickle cell disease in order to help prevent serious problems with your health. Every person with sickle cell disease should have a hematologist and a primary care physician that knows the patient's detailed medical history in order to provide the best medical care possible.¹

It is critical that people with sickle cell disease stay hydrated at all times. Drinking 8 glasses of water each day is a good way to keep the body hydrated. Eating a balanced meal and making healthy choices are important for proper nutrition.¹

Maintaining a balanced body temperature is very important, getting too hot or too cold can cause sickness. Be sure to wear layers to adapt to changing temperatures.¹

In general, people who have sickle cell disease have a reduced life expectancy. Some people with the disease can remain without symptoms for years, while others do not survive beyond infancy or early childhood. New treatments for sickle cell disease are improving life expectancy and quality of life. People with sickle cell disease can survive beyond their 50s with optimal management of the disease.¹

INHERITENCE PATTERN OF SICKLE CELL DISEASE

A simple blood test is used to diagnose SCD and is often during routine newborn screening tests at the hospital however, SCD can be diagnosed before birth. Early diagnosis and treatment is important as children with SCD are at an increased risk of infection and other health problems. Common types of SCD include:¹³

- **HbSS** is the most severe form of SCD, occurring in those who inherit one sickle cell gene ('S') from each parent.
- **HbSC** is a milder form of SCD and is found in those who inherit a sickle cell gene ('S') from one parent and a gene for an abnormal hemoglobin called 'C' from the other parents.
- **HbS beta thalassemia** occurs when one inherit one sickle cell gene ('S') from one parent and one gene for beta thalassemia from the other parent. There are two types of beta thalassemia: '0' and '+'. Individuals with *HbS beta 0-thalassemia* usually have a severe form of SCD; those with *HbS beta +-thalassemia* may have a milder form.
- **HbSD, HbSE, and HbSO** are rare types of SCD and are found in those who inherit one sickle cell gene ('S') and one gene from an abnormal type of hemoglobin ('D', 'E', or 'O').

HbAS is found in individuals who have a sickle cell trait and inherit one sickle cell gene ('S') from one parent and one normal gene ('A') from the other parent. This is also known as sickle cell trait (SCT) – individuals with SCT usually do not have any of the signs of the disease and live a normal life, but can pass the trait on to their children.¹³

- If both parents have SCT, there is a 50% chance that any child of theirs also will have SCT. Such children will not have symptoms of SCD, but they can pass SCT on to their children.
- If both parents have SCT, there is a 25% (or 1 in 4) chance that any child of theirs will have SCD. There is the same 25% (or 1 in 4) chance that the child will not have SCD or SCT.
- If one parent has SCT, there is a 50% (or 1 in 2) chance that any child of this parent will have SCT and an equal 50% chance that the child will not have SCT.