Closing Gaps & Meeting Metrics

Coding Tips & Best Practices

June 2023

Sickle Cell Disease

June 19 is World Sickle Cell Day, an annual observance to increase public knowledge and understanding of sickle cell disease, a life-long illness. While the exact number of people living with sickle cell disease in the United States is unknown, it is estimated that 100,000 Americans are affected by the disease.

Sickle cell disease is a group of inherited red blood cell disorders that affect hemoglobin, the protein that carries oxygen through the body. Normally, red blood cells are disc-shaped and flexible enough to move easily through the blood vessels. With sickle cell disease, the red blood cells are crescent- or "sickle"-shaped. These cells do not bend or move easily and can block blood flow in the body.

Sickle cell disease affects diverse groups of people. Experts agree that it is important to get tested, especially if you have family members who have been diagnosed with sickle cell disease. The Centers for Disease Control (CDC) has helpful resources online to learn more about this disease.

How Sickle Cell Disease May Affect Your Health

Sickle cell disease can affect people in different ways. Some of these are:

- Acute pain crisis: Also known as sickle cell or vaso-occlusive crisis, this can happen without warning when sickle cells block blood flow.
- Chronic (long-term) pain: Chronic pain is common, but it can be hard to describe. It is usually different from crisis pain or the pain that results from organ damage.
- Delayed growth and puberty: Because of anemia, children who have sickle cell disease may grow and develop more slowly than their peers.
- Infections: In sickle cell disease, a damaged spleen raises the risk for certain infections, including chlamydia, Haemophilus influenzae type B, salmonella, and staphylococcus.
- Joint problems: Sickling in joints such as hips, shoulders, knees, and ankles can lower oxygen flow and result in a condition called avascular or aseptic necrosis, causing severe damage to the joints.
- Pregnancy problems: Pregnancy can raise the risk of high blood pressure and blood clots in people with sickle cell disease. The condition also increases the risk of miscarriage, premature birth, and low birth-weight babies.

Symptoms

- A yellowish color of the skin (jaundice) or whites of the eyes (icterus) that occurs when many red cells undergo hemolysis
- Extreme tiredness or fussiness from anemia
- Painful swelling of the hands and feet, known as dactylitis

Diagnosis Methods

- Blood test and genetic tests
- Prenatal screening
- Newborn screening

Treatments

- Medicine to prevent the sickling of red blood cells
- Medicine to reduce vaso-occlusive and pain crises
- Medicine to treat pain
- Medicine to reduce the risk of infection
- Blood transfusions
- Blood or bone marrow transplant
- Potential genetic therapy treatments: The National Heart, Lung, and Blood Institute (NHLBI) is exploring ways genetic therapies may help provide new treatments or cure for sickle cell disease. Genetic therapies aim to treat or cure conditions by adding new DNA or changing existing DNA.

Documentation Tips

- Document the purpose of the encounter, (e.g., Follow up visit for treatment and evaluation of…)
- Identify the type of sickle cell disease (e.g., Hb-C, spherocytosis, thalassemia, etc.)
- Document specific diagnosis, not just abnormal laboratory values. Coders cannot abstract sickle cell disease based on laboratory values reported in the record.
- Capture the complications and associated conditions (e.g., acute chest syndrome, enlargement of the spleen, etc.)
- Include positive physical findings or indicators (e.g., pain, vision loss, etc.)
- Describe each final diagnosis to the highest level of specificity (e.g., sickle cell thalassemia with cerebral vascular involvement, etc.)
- Document the patient's blood or bone marrow status if the patient has had a bone/blood marrow-transplant
- SCD is a common abbreviation for sickle cell disease. SCD is also used to describe other medical conditions such as spinal cord disease. To avoid confusion, always spell out the first reference to an abbreviation. (e.g., sickle cell disease (SCD).
- Document a clear and concise treatment plan.

Coding Tips

In coding sickle cell disorders, it is important to understand the difference between sickle cell anemia or disease and sickle cell trait. Sickle cell trait occurs when a child receives the genetic trait from only one parent. People with sickle cell trait typically do not develop sickle cell disease. Code this as D57.3 Sickle-cell trait.

When the medical record contains both the terms *sickle cell trait* and *sickle cell disease*, capture only the code for sickle cell disease (D57.0-, D57.2-, D57.4, and D57.8-)

Sickle Cell Disease Coding Reference

Code	Description	Includes
D57.0-	Hb-SS disease with crisis	
D57.1	Sickle-cell disease without crisis	Hb-SS disease without crisis Sickle-cell anemia NOS Sickle-cell disease NOS Sickle-cell disorder NOS
D57.2-	Sickle-cell/Hb-C disease	Hb-SC disease Hb-S/Hb-C disease
D57.3	Sickle-cell trait	Hb-S trait Heterozygous hemoglobin S
D57.4-	Sickle-cell thalassemia	Sickle-cell beta thalassemia Thalassemia Hb-S disease
D57.8-	Other sickle-cell disorders	Hb-SD disease Hb-SE disease

References

- Sickle Cell Disease - What Is Sickle Cell Disease? | NHLBI, NIH
- Learn More About Sickle Cell Disease | CDC
- https://www.encoderpro.com/epro/index.jsp
- https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm