SEPTEMBER 2025

Sickle Cell Disorders

At Healthfirst, we are committed to helping providers accurately document and code their patients' health records. This tip sheet is intended to assist providers and coding staff with the documentation and ICD-10-CM selection, on services submitted to Healthfirst—specifically for **Sickle Cell Disorders**. It provides information from industry sources about proper coding practice. However, this document does not represent or guarantee that Healthfirst will cover and/or pay for the services outlined. Coverage decisions are based on the terms of the applicable evidence of coverage and the provider's participation agreement. This includes the determination of any amount that Healthfirst or the member owes the provider.

Sickle Cell Disorders are severe, chronic diseases caused by a genetic variation in the hemoglobin protein in the red blood cell.

Diagnosis and Descriptions

ICD-10-CM	Description
D57.00	Hb-SS disease with crisis, unspecified
D57.01	Hb-SS disease with acute chest syndrome
D57.02	Hb-SS disease with splenic sequestration
D57.03	Hb-SS disease with cerebral vascular involvement
D57.04	Hb-SS disease with dactylitis
D57.09	Hb-SS disease with crisis with other specified complication
D57.1	Sickle-cell disease without crisis
D57.20	Sickle-cell/Hb-C disease without crisis
D57.21*	Sickle-cell/Hb-C disease with crisis
D57.3	Sickle-cell trait Includes: Hb-S trait, Heterozygous Hb-S
D57.40	Sickle-cell thalassemia without crisis
D57.41*	Sickle-cell thalassemia, unspecified, with crisis

ICD-10-CM	Description
D57.42	Sickle-cell thalassemia beta zero without crisis
D57.43*	Sickle-cell thalassemia beta zero with crisis
D57.44	Sickle-cell thalassemia beta plus without crisis
D57.45*	Sickle-cell thalassemia beta plus with crisis
D57.80	Other sickle-cell disorders without crisis
D57.81*	Other sickle-cell disorders with crisis

**6th characters identified as 1 - acute chest syndrome, 2 - splenic sequestration, 3 - cerebral vascular involvement, 4 – dactylitis, 8 - other specified complication, 9 – with crisis NOS

Coding Tips:

- Code also, if applicable, cerebral infarction (I63.0-I63.9) for code D57.03
- Use additional code to identify complications, Such as:
  - cholelithiasis (K80-K80.81) priapism (N48.32) for code D57.09
- D57.00 - Hb-SS disease with crisis, unspecified, Include terms: -With (painful) crisis NOS, -With (vaso-occlusive) pain NOS
- D57.1 - Sickle-cell disease without crisis, Include terms: Sickle-cell anemia NOS, Sickle-cell disease NOS, Sickle-cell disorder NOS
- D57.41* - Sickle-cell thalassemia, unspecified, with crisis, Include terms: Microdrepanocytosis, Sickle-cell thalassemia NOS.

Documentation Recommendations

Causes	Types of Sickle Cell Disorder	Diagnostic Tests	Complications

• Genetic
• Inherited
	• HbSS
	• HbSC
	• HbSD
	• HbSE
	• HbSO
	• Sickle Cell Traits
	  ○ HbAS
	• HbS Beta Thalassemia
	  ○ Thalassemia beta zero
	  ○ Thalassemia beta plus
	• Sickle-cell retinopathy
	  ○ (H36.811 - H36.819, H36.821 - H36.829)
		• Chorionic Villus Sampling
		• Amniocentesis
		• Routine Newborn Screening Tests
			• Acute Chest Syndrome
			• Dactylitis
			• Splenic Sequestration
			• Priapism
			• Liver Problems
			• Fever/Infection
			• Anemia
			• Blood Clots
			• Stroke
			• Organ Damage
			• Avascular Necrosis
			• Kidney Problems
			• Leg Ulcers
			• Pulmonary Hypertension

Treatment			
Lifestyle Behaviors	Prevention of Infections, Anemia and Stroke	Management of Pain Crises	Treatment for SCD
• Drink Plenty of Water • Avoid getting too hot or too cold • Avoid places or situations that cause Exposure to High Altitudes	• Wash your hands often • Vaccines • Transcranial Doppler Ultrasound	• Intravenous Fluids • Pain-Reducing Medicine • Hospitalization for severe pain crises	• Bone Marrow or Stem Cell Transplants • Blood Transfusion • Hydroxyuria • Splenectomy

Coding Example:

Case 1	A six-month-old infant with sickle-cell anemia presented with fever and puffy tender feet and hands. The provider diagnosed SCD with dactylitis. How is this diagnosis coded?
AHA Coding Clinic 2023 Fourth Quarter Rationale	Assign code **D57.04** - Hb-SS disease with dactylitis, for SCD with dactylitis. Assign code **R50.81** - Fever presenting with conditions classified elsewhere, as an additional diagnosis.

References

- ICD-10 Coding Guidelines
- Coding Clinic Advisor
- Sickle Cell Disease | Merck Manuals

Questions?

Contact us at #Risk_Adjustments_and_clinical_Documentation@healthfirst.org.

For additional documentation and coding guidance, please visit the coding section on HFproviders.org