Chapter 3: Diseases of the Blood & Blood-Forming Organs and Certain Disorders Involving the Immune Mechanism

Sickle-cell disorders

Code Axes

- Hb-SS disease with crisis: D57.0-
- Sickle-cell disease without crisis: D57.1
- Sickle-cell/Hb-C disease: D57.2-
- Sickle-cell trait: D57.3
- Sickle-cell thalassemia: D57.4-
- Other sickle-cell disorders: D57.8-

I-10 Alert:

When the medical record documentation indicates fever is present code R50.81 should also be reported.

Translation

The ICD-10-CM subcategories for sickle-cell disorders include combination codes representing associated conditions, such as acute chest syndrome and splenic sequestration. The condition is a group of red blood cell disorders in which the cells become hard, clump together, and the resulting C-shape resembles a sickle. These cells die early, causing a constant shortage of healthy red blood cells.

Hb-SS disease with and without crisis (D57.0-, D57.1)

Hb-SS disease is assigned for patients who inherited two sickle-cell hemoglobin genes (“S”), one from each parent. This is the most severe form of the disease, is chronic and often incurable, and in many cases requires multiple transfusions. The sickle-cell crisis refers to recurring acute episodes of pain involving any body system, but usually the chest, bones, or abdomen. The sickle-shaped cells may “clog” the blood vessels, causing pain and other problems.

Acute chest syndrome: a vasoocclusive crisis involving the pulmonary vasculature. It’s typically caused by an underlying lung infection, which then leads to further inflammation, red-blood cell sickling with further vasoocclusion, and a drop in oxygen levels. The condition is life-threatening and is one of most common causes of death in the SCD patient.
Splenic sequestration: When the blood vessels leading to and from the spleen become obstructed due to sickle-shaped red blood cells, the blood stays in the spleen, causing enlargement (splenomegaly) and in some cases, a precipitous drop in red blood cell count. Recurrent episodes are common, necessitating multiple transfusions and possibly splenectomy. Infants and pediatric patients are at a higher risk for splenic sequestration.

Key Terms
- Key terms found in the documentation may include:
  - Sickle-cell anemia
  - Sickle-cell disease
  - Sickle-cell disorder
  - Hb-SS disease with vasoocclusive pain
  - Sickle-cell disease with crisis
  - Hb-SS with acute chest syndrome
  - Hb-SS with splenic sequestration
  - drepanocytosis
  - hemolytic crisis

CDI Alert
Documentation for patients with SCD and symptoms including sudden weakness, pain on left side of abdomen, tachycardia, or a new infiltrate on chest x-ray may reflect one of the more severe forms of the disease, such as acute chest syndrome or splenic sequestration.

Hospital Alert
Codes in this section for SCD with crisis (D57.00, D57.01, D57.02) are designated as Major Complication / Comorbidities (MCC), reflecting the highest severity level.