ICD-10-CM Documentation and Coding Best Practices
Sickle Cell Disease

Overview

Sickle cell disease, also known as sickle cell anemia, is a blood disorder in which the red blood cells are sickle shaped and are unable to move through blood vessels smoothly. They have shorter life span because they are more fragile, causing a deficiency in hemoglobin – the protein in red blood cells that carries oxygen throughout the body.

Cause

- Homozygous inheritance of the Hbs-producing gene (defective Hb gene from each parent)

What can cause a sickling episode? At first, sickling is reversible with oxygenation but eventually becomes irreversible due to cell membrane damage.

- Infection
- Stress-emotional or exertional physical
- Surgery
- Dehydration
- Acidosis
- Cold
- Anything that leads to hypoxia

Symptoms

- Pain in back, extremities, chest, and abdomen during sickling episode (can be moderate-severe)
- Fever, swilling of hands/feet, tenderness
- Tachypnea
- Hypertension
- Nausea/vomiting
- Priapism

Symptoms of advanced sickle cell disease (due to repeated occlusion of vessels)

- Splenomegaly, hepatomegaly, renal insufficiency, gallstones
- Cardio: heart failure, MI, HTN, cor pulmonale
- Eyes: retinal detachment, scarring
- Skin: pallor, fatigue, jaundice, skin ulcer, esp. of legs
- Bone: deformity and fractures due to thinning and flattening

Types of Sickle cell disease

- Hemoglobin SS disease
- Hemoglobin SC disease
- Hemoglobin SB+ (beta) thalassemia
- Hemoglobin SB 0 (Beta-zero) thalassemia
- Hemoglobin SD, hemoglobin SE, and hemoglobin SO
- Sickle cell trait

Treatment

- Oxygenation
- Fluid replacement to thin blood-ORAL & IV
- Rest – but may need anticoagulants to prevent DVT
- Blood transfusions – but if this causes iron overload, give Desferal (deferoxamine)
- Erythrocytapheresis – exchange of HGBs to regular HGB (like dialysis, but just RBC exchange)

Medications

- Analgesics – morphine/NSAID
- Hydroxyurea – decreases production of abnormal cells for pts. w/frequent crisis episodes (need frequent serum studies since it is chemo that suppresses bone marrow)
- Penicillin – decrease risk of trigger due to sickness (may be prophylactic prior to med/dental procedures)
- Prophylactic antibiotics (reduce infection risk)
- Folic acid replacement – helps to increase HGB levels and helps with hemolysis

Coding and Documentation Guidance

- Document type of disease
- Document with or without crisis
- Identify the crisis
    - Acute chest syndrome
    - Splenic sequestration

ICD-10-CM Codes

- D57.0-Hb SS disease with crisis
    - D57.00-Hb-SS disease with crisis, unspecified
    - D57.01-Hb-SSS disease with acute chest syndrome
    - D57.02-Hb-SSS disease with splenic sequestration
- D57.1-Sickle-cell disease without crisis
- D57.2-Sickle-cell/Hb-C disease
    - D57.20-Sickle-cell/Hb-C disease without crisis
    - D57.21-Sickle-cell/Hb-C disease with crisis
        - D57.211-Sickle-cell/Hb-C disease with acute chest syndrome
        - D57.212-Sickle-cell/Hb-C disease with splenic sequestration
        - D57.219-Sickle-cell/HB-C disease with crisis, unspecified
- D57.3-Sickle-cell trait
- D57.4-Sickle-cell Thalassemia
    - D57.40-Sickle-cell thalassemia without crisis
    - D57.41-Sickle-cell thalassemia with crisis
        - D57.411-Sickle-cell thalassemia with acute chest syndrome
        - D57.412-Sickle-cell thalassemia with splenic sequestration
        - D57.419-Sickle-cell thalassemia with crisis, unspecified
- D57.8-Other sickle-cell disorders
    - D57.80-Other sickle-cell disorders without crisis
    - D57.81-Other sickle-cell disorders with crisis
        - D57.811-Other sickle-cell disorders with acute chest syndrome
        - D57.812-Other sickle-cell disorders with splenic sequestration
        - D57.819-Other sickle-cell disorders with crisis, unspecified