AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Sickle Cell Anemia

Quick Reference Guide

2026

Table of Contents

1 CLINICAL SNAPSHOT

Definition: Sickle cell disease (**SCD**) is a group of inherited red blood cell disorders caused by autosomal-recessive mutations in the **HBB gene** that produce **abnormal hemoglobin S** (**HbS**).[1,2] Under deoxygenated conditions, **HbS polymerizes** into rigid rods that **distort red cells into a sickle shape**, reducing red cell lifespan from **120 days** to **10–20 days** and obstructing microvascular flow; this drives a cascade of vasculopathy, hemolytic anemia, acute pain crises, end-organ damage, and premature death.[1,2] SCD is a **spectrum of genotype-defined disorders**: **HbSS** (sickle cell anemia, most severe), **HbS/β^0-thalassemia** (functionally equivalent to HbSS), **HbSC** disease (generally milder but still causes significant complications), **HbS/β$^+$-thalassemia** (variable severity), and rare variants **HbSD**, **HbSE**, and **HbSO-Arab**.[2,3] **Sickle cell trait** (HbAS, one normal HBB allele) is a **carrier state, NOT active disease**.[3,4]

ICD-10 Codes:

D57.x designates sickle-cell disorders, categorized by genotype, crisis status, and specific complication.[5] Common codes: **D57.00** (HbSS with crisis, unspecified), **D57.01** (HbSS with acute chest syndrome (ACS) = **leading cause of SCD death**), **D57.02** (HbSS with splenic sequestration), **D57.03** (HbSS with cerebral vascular involvement), **D57.1** (sickle-cell disease without crisis; **genotype-nonspecific**), **D57.20** (HbSC without crisis), **D57.211** (HbSC with ACS), **D57.42** (HbS/β^0-thal without crisis), **D57.44** (HbS/β$^+$-thal without crisis), and **D57.3** (sickle-cell TRAIT; **carrier state only**). Complication codes paired alongside **D57**: **N18.x** (CKD), **I27.2x** (secondary pulmonary hypertension), **H35.x** (proliferative sickle retinopathy), **M87.x** (avascular necrosis), **F32.x** (depression).

Prevalence and Burden: SCD affects approximately **100,000 people** in the United States, predominantly **Black Americans** (**1 in 365** Black births) and **Hispanic Americans** (**1 in 16,300** Hispanic births).[6,7] More than 3 million Americans carry **sickle cell trait** (**D57.3**).[8] Life expectancy for publicly insured individuals with SCD is approximately **52.6 years**, roughly **20 years shorter** than the general population, meaning few patients historically survived to age 65.[2,9] **Hydroxyurea therapy**, infection prophylaxis, and comprehensive care are now extending survival, with median survival increasing from age 14 years in 1973 to **as high as 61 years** in recent academic center cohorts, creating a growing but heavily comorbid Medicare-age SCD population.[10,11] Adult Medicaid enrollees with SCD incur mean total costs of **$45,114/year** vs. **$6,039** for matched non-SCD adults (a 7.5-fold difference), driven primarily by hospitalizations (18.7-fold higher costs) and ED visits (12.7-fold higher costs).[12] Estimated lifetime medical costs reach **$1.6–$1.7 million** per patient.[12] Approximately **222,612 ED visits** per year are attributable to SCD nationally, three-fourths for **pain**.[13] Medicare-specific PMPY data for adults aged 65+ are **essentially absent**: the Medicaid figures are the best available proxy, though the Jiao et al. nationwide cohort study of 94,616 Medicare and Medicaid beneficiaries demonstrated **significant survival disparities by insurance type** even among those reaching age ≥65.[9,12]