AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Sideroblastic Anemia

Quick Reference Guide

2026

Table of Contents

1 CLINICAL SNAPSHOT

Definition: **Sideroblastic anemia** (SA) is a heterogeneous group of disorders **unified by one marrow finding**: **ring sideroblasts** = erythroid precursors with at least **five** siderotic granules covering a minimum of **one-third** of the nuclear circumference[2] on Prussian blue (Perls) staining.[1,2] Iron accumulates in the mitochondria of developing red cells because it **cannot be incorporated into hemoglobin**; the body has iron, but the **machinery to use it is impaired**.[3,4] SA is classified as **congenital** (most often X-linked, from ALAS2 mutations, predominantly affecting males and typically microcytic) or **acquired**.[1,5] Acquired forms span **a spectrum from reversible drug-, toxin-**, and **nutrition-related disease** to clonal hematologic malignancy - most importantly **myelodysplastic syndrome with ring sideroblasts** (MDS-RS), the dominant acquired form in older adults, driven by SF3B1 somatic mutations in approximately **80%** of cases.[1,6] Red-cell size **varies by subtype**: congenital SA is typically **microcytic**, while acquired MDS-RS is frequently **normocytic or macrocytic**; **do not** use **MCV** alone to exclude SA.[7,8]

ICD-10 Codes

Sideroblastic anemia is reported with **etiology-specific subcodes**: **D64.0** (hereditary SA), **D64.1** (secondary SA **due to disease**), and **D64.3** (other SA, including pyridoxine-responsive and SA not otherwise specified) map to HCC 109; **D64.2** (secondary SA due to drugs and toxins) also maps to HCC 109.[9] When a bone marrow study confirms **MDS with ring sideroblasts**, the correct codes are **D46.1** or **D46.B**. **D64.9** (anemia, unspecified) should be avoided once ring sideroblasts are confirmed.[9] Code **reversible drivers** and **complications concurrently**: **F10.20** (alcohol use disorder), **E61.0** (copper deficiency), **E53.1** (vitamin B6 deficiency), **T56.0-** (lead toxicity), and **E83.10-E83.19/E83.111** (iron overload/transfusion-related hemochromatosis).[9]

Prevalence and Burden: No US population-based prevalence study exists for sideroblastic anemia (**D64.0-D64.3**) as a standalone category; all figures below are **MDS-derived proxy data** and should be read as such.[4,10] MDS incidence is approximately **4 per 100,000** persons per year in the United States, rising to about **25 per 100,000** in persons aged 65 and older; some estimates using SEER-Medicare claims-based algorithms put the true rate near **75 per 100,000** among persons 65 and older due to **registry underreporting**.[10,11] A 2026 update places the median age at MDS diagnosis at **76 years** among US patients.[12] **More than 10,000** MDS diagnoses occur annually in the United States, with approximately **86%** in individuals aged 60 and older.[12,13] The ring-sideroblast subtype accounts for roughly **4–10% of MDS cases**, and **SF3B1-mutant MDS-RS** (the most common genetically defined MDS subtype) carries a relatively favorable median survival of about **6.6 years** in Mayo Clinic data.[1,14] **Anemia in older adults** independently **predicts mortality, hospitalization, falls, and functional decline regardless of its cause**.[15]