



# **Sideroblastic Anemia**

## **Quick Reference Guide**

2026

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# 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<sup>2</sup> on Prussian blue (Perls) staining.<sup>1,2</sup> 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**.<sup>3,4</sup> SA is classified as **congenital** (most often X-linked, from ALAS2 mutations, predominantly affecting males and typically microcytic) or **acquired**.<sup>1,5</sup> 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.<sup>1,6</sup> 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.<sup>7,8</sup>

## 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.<sup>9</sup> 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.<sup>9</sup> 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).<sup>9</sup>

**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.<sup>4,10</sup> 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**.<sup>10,11</sup> A 2026 update places the median age at MDS diagnosis at **76 years** among US patients.<sup>12</sup> **More than 10,000** MDS diagnoses occur annually in the United States, with approximately **86%** in individuals aged 60 and older.<sup>12,13</sup> 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.<sup>1,14</sup> **Anemia in older adults** independently **predicts mortality, hospitalization, falls, and functional decline regardless of its cause**.<sup>15</sup>

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