AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Hemophilia

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Hemophilia is a **hereditary X-linked recessive bleeding disorder** caused by deficiency of coagulation factor VIII (Hemophilia A) or factor IX (Hemophilia B); rarer factor XI deficiency is Hemophilia C.[1] Severity is classified by baseline factor activity level: severe (<1%) — spontaneous joint and muscle bleeds; moderate (1–5%) — bleeding after minor trauma; mild (5-40%) — bleeding only after surgery/major trauma.[2] **Acquired Hemophilia A (AHA) is clinically distinct** — autoimmune inhibitory antibodies against endogenous FVIII, median age at diagnosis around 75 years, incidence 3–4 per million/year in adults ≥60, mortality ≥20% in patients >65.[3]

ICD-10 Codes:

Hereditary: **D66** (Hemophilia A, FVIII deficiency); **D67** (Hemophilia B, FIX deficiency); **D68.1** (Hemophilia C, FXI deficiency). Acquired: **D68.311** (Acquired Hemophilia A, autoimmune inhibitors) — clinically and codewise distinct from hereditary; **D68.4** (acquired factor deficiency, NOT autoimmune) does not map to HCC. **AVOID: Z14.01** (asymptomatic carrier) and **Z14.02** (symptomatic carrier) carry NO HCC weight — never use as primary for active hemophilia disease. Associated bleeding codes: **M25.0x** (hemarthrosis by joint, ~70–80% of bleeds), **I61.x** (intracranial hemorrhage), **K92.0–K92.2** (GI hemorrhage), **M17.4–M17.5** (hemophilic arthropathy, secondary OA), **B18.2** (chronic HCV — ~80% seroprevalence in pre-1990 cohort), **C22.0** (hepatocellular carcinoma surveillance), **I48.x** (atrial fibrillation), **I25.x** (ischemic heart disease).

Prevalence and Burden: The estimated frequency of hemophilia is around **1 in 10,000 live births**.[1] Hemophilia A affects **~1 in 5,000** male births (~20,000 US persons); Hemophilia B **~1 in 30,000** male births (~4,000 US persons);[1] ~30% of all cases arise from de novo mutations with **no family history**.[4] Inhibitor development is the most costly complication: 30–50% of severe Hemophilia A patients develop FVIII inhibitors; Hemophilia B rate 1.5–3%.[5,6] Clotting factor concentrates account for ~90% of total direct medical costs in PwH treated at HTCs.[7]

HCC/RAF V28 Mapping

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
D66 (Hemophilia A, FVIII)	HCC 111 — Hemophilia/Male	4.639	Hereditary etiology confirmed; baseline FVIII activity level (% or IU/mL); inhibitor status (positive/ negative, titer in Bethesda Units); current prophylaxis regimen; documented annually in MA (not a carry-forward diagnosis)[7]
D67 (Hemophilia B, FIX)	HCC 111	4.639	Hereditary etiology; baseline FIX activity level; inhibitor status; note: emicizumab is NOT indicated for Hem B — document correct type to support prescribing[7]