AAVBC

AMERICAN ACADEMY OF VALUE BASED CARE

ALS

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Amyotrophic lateral sclerosis (ALS) — also called Lou Gehrig disease — is a progressive, uniformly fatal neurodegenerative disease that destroys **upper motor neurons (UMN)** in the brain and **lower motor neurons (LMN)** in the brainstem and spinal cord, producing relentless **muscle atrophy, paralysis, and ultimately respiratory failure**.[1] Median survival from symptom onset is **2-5 years**, and **respiratory failure is the cause of death in up to 95%** of people living with ALS.[2] Approximately **90-95% of cases are sporadic** (unknown cause); **5-10% are familial**, with pathogenic variants in C9orf72, SOD1, TARDBP, and FUS among the most characterized.[1] ALS is not purely a motor disease: up to **50% of patients develop cognitive or behavioral changes**, and roughly **15% develop frontotemporal dementia (ALS-FTD)** — a finding routinely missed in primary care with profound implications for advance care planning and decision-making capacity.[3]

ICD-10 Codes:

Primary diagnosis: **G12.21** — Amyotrophic lateral sclerosis — is the specific code for confirmed ALS and should be documented whenever the phenotype is known. Document "amyotrophic lateral sclerosis" or "ALS" explicitly; both familial and sporadic ALS map to **G12.21** (there is no separate code), so record genetic testing results (SOD1, C9orf72, FUS, TARDBP) in the chart to support specialist referral and **tofersen** eligibility. Avoid **G12.20** (motor neuron disease, unspecified) once the ALS phenotype is confirmed — it was the default before ICD-10 carried a specific ALS code and forfeits clinical specificity. Related motor neuron disease codes (**G12.22** progressive bulbar palsy; **G12.23** primary lateral sclerosis; **G12.25** progressive spinal muscular atrophy) describe distinct phenotypes and should not substitute for **G12.21** when ALS is confirmed. Because ALS progresses rapidly, comorbidity codes for respiratory, nutritional, mobility, and cognitive status must be updated at every encounter.[4]

Prevalence and Burden: The CDC National ALS Registry estimated approximately **33,000 people living with ALS** in the United States in 2022 (prevalence **~9.9 per 100,000**), projected to rise more than 10% to **over 36,000 by 2030** as the population ages; in the Medicare Advantage population specifically, 5-year period prevalence has been reported at **20.5 per 100,000**.[5,6] ALS incidence peaks between ages **55 and 75 years**, with a male-to-female ratio of approximately **1.2:1 to 1.5:1** for sporadic disease (1:1 for familial). The older population (>65 years) carries a disproportionate burden: in beneficiaries **≥65 years**, median time to diagnosis is **2.5 years for limb-onset** and **1.25 years for bulbar-onset**, and roughly **40% of Medicare ALS patients died within 1 year** of their first ALS diagnostic code or never saw a neurologist — with **25.5% dying within 6 months** — evidence of severe under-ascertainment in older adults.[1]