



Muscular Dystrophy

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Muscular dystrophy (MD) is a group of genetically determined myopathies causing **progressive skeletal-muscle degeneration and weakness**, driven by mutations in the **dystrophin-glycoprotein complex** and related structural, enzymatic, and nuclear-envelope proteins.¹ More than **40 causative genes** are recognized across subtypes.² Although MD is widely stereotyped as a pediatric disease, several forms present or accelerate in older adults - **myotonic dystrophy type 2 (DM2)**, **limb-girdle MD (LGMD)**, **facioscapulohumeral MD (FSHD)**, and **oculopharyngeal MD (OPMD)** - where adult-onset diagnostic delay runs about **4 to 14 years**.³⁻⁷ The clinical stakes in older adults are **systemic**, not just motor: cardiomyopathy and conduction disease, restrictive respiratory failure, and dysphagia with aspiration risk all track with these disorders.

ICD-10 Codes:

MD is coded under category **G71.0-**, and the FY2026 tabular provides granularity down to the specific protein-dysfunction level - always assign the most specific code the record supports. **G71.01** covers Duchenne or Becker MD; **G71.02** facioscapulohumeral MD; the LGMD series runs **G71.031** (autosomal dominant), **G71.032** (calpain-3), **G71.033** (dysferlin), **G71.0340-.0349** (sarcoglycan, 6th character = alpha/beta/other), **G71.035** (anoctamin-5), and **G71.036** (fukutin-related protein); **G71.09** documents other specified MD - **Emery-Dreifuss**, **OPMD**, and **congenital MD** (which has no dedicated code). Myotonic MD is **G71.11**. Reserve **G71.00 (unspecified)** for cases genuinely pending workup - it lacks specificity and invites audit scrutiny.⁸

Prevalence and Burden: MDs are individually rare and reported by subtype rather than in aggregate. **Myotonic dystrophy is the most common adult-onset MD**, with an estimated incidence of **~1 per 8,000** and adult DM1 prevalence **~10.6 per 100,000**.^{2,9} DM2 is likely underestimated, with a diagnostic delay averaging **14 years**.⁴ **FSHD** affects **~3 per 100,000**,² and **LGMD** ranges from **1:14,500¹⁰ to 1.63:100,000¹¹** by subtype. DMD/BMD incidence is **~1 in 5,000-6,000 live male births**, about one-third from de novo mutations.^{12,13}

HCC/RAF V28 Mapping

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
G71.0- (Muscular dystrophy, all subcodes)	HCC 197 - Muscular Dystrophy	0.426	Named subtype + genetic and/or CK evidence; confirm at least annually
G71.11 (Myotonic disorders)	HCC 197 - Muscular Dystrophy	0.426	Specify DM1 vs DM2; CTG/CCTG repeat expansion
G71.20-.29- (Congenital myopathies)	HCC 197 - Muscular Dystrophy	0.426	Named myopathy (nemaline, myotubular, centronuclear)

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