AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

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 **1:14,500**[10] **to 1.63:100,000**[11] 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)

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT

ABBREVIATIONS: ABBREVIATIONS: HCC = Hierarchical Condition Category; RAF = Risk Adjustment Factor; CNA = Community Non-Dual Eligible, Aged; MD = muscular dystrophy; DM1/DM2 = myotonic dystrophy type 1/2. *Annual value = RAF 0.426 x $10,402.34 CY2026 MA base rate. *RAF values represent the Community Non-Dual Eligible Aged (CNA) coefficient from the 2026 CMS-HCC model; values vary across patient populations based on eligibility and care setting

Risk-Adjusted Care Resources per Patient/Year[14,15]

Risk-adjusted care resource allocation — MA base rate (~$10,402.34) × RAF coefficient

Muscular Dystrophy
~$4,431

HCC 197 · RAF 0.426

2 RECOGNITION AND DIAGNOSIS

Medicare Screenings and Diagnostic Coverage

TEST	COVERAGE	FREQUENCY	CPT	CLINICAL INDICATION
Creatine kinase (CK), total[2]	Part B diagnostic lab	As medically necessary	82550	**First-line** when unexplained weakness/falls; note normal CK does **NOT** exclude late-onset MD
Spirometry (upright + supine)[16,17]	Part B	At diagnosis, then periodically	94010	**Restrictive disease surveillance**; PFTs are the only reliable pre-symptom detection
Echocardiogram, complete TTE[10,18]	Part B	At diagnosis, then annually (at-risk subtypes)	93306	**Cardiomyopathy surveillance** in DMD/BMD, LGMD, sarcoglycanopathy, DM1/DM2
ECG, 12-lead[18]	Part B	Annually (DM1/DM2, EDMD, LGMD1B)	93000	**Conduction disease**; laminopathies carry sudden-death risk

TEST	COVERAGE	FREQUENCY	CPT	CLINICAL INDICATION
Needle EMG (2 extremities)[19]	Part B	Diagnostic workup	95861	**Myotonic discharges support DM1/DM2**; low yield for subtype otherwise
Genetic testing/ NMD panel[20,21]	Coverage varies by MAC; PA often required	Diagnostic	81408/ 81479	**Confirms subtype**; first-line for distinctive phenotypes (DM, FSHD, OPMD)
Swallowing evaluation[17,22]	Part B	When dysphagia suspected	92610	OPMD, DM1/DM2, advanced MD; **aspiration-risk assessment**

ABBREVIATIONS: ABBREVIATIONS: CK = creatine kinase; TTE = transthoracic echocardiogram; PFT = pulmonary function test; EMG = electromyography; NMD = neuromuscular disease; MAC = Medicare Administrative Contractor; PA = prior authorization; CPT = Current Procedural Terminology

Subtle Early Signs in Older Adults (>65)

SUBTLE EARLY SIGN (>65)	WHY IT MATTERS/COMMONLY MISATTRIBUTED TO
Asymmetric muscle wasting (one leg/ shoulder wasting faster)[23,24]	**Asymmetry does not occur in normal sarcopenia**; warrants CK and neurology referral
Difficulty rising from a chair; waddling gait; frequent tripping[2,25]	**Proximal (hip/shoulder-girdle) weakness** routinely chalked up to sarcopenia or general frailty rather than dystrophic wasting
Proximal weakness with hip/back pain[24,26]	**Misattributed to osteoarthritis, spinal stenosis, or lumbosacral radiculopathy** -> orthopedic referral, missing the myopathy
Progressive dysphagia and/or ptosis[27]	In OPMD, **mistaken for silent stroke, GERD, esophageal dysmotility, or myasthenia gravis**
Myalgia/weakness in a patient on a statin[28]	Reflexively called statin myopathy; **statins can unmask or aggravate subclinical late-onset MD**
Atypical cardiac (arrhythmia, conduction block) or metabolic (atypical diabetes) presentation[18,29,30]	Late-onset LGMD and myotonic dystrophy can present with **organ involvement before overt limb weakness**

SUBTLE EARLY SIGN (>65)	WHY IT MATTERS/COMMONLY MISATTRIBUTED TO
Weakness that fails to improve with a course of physical therapy[31]	**Persistent non-improvement** warrants EMG or targeted genetic panel rather than continued rehab alone

ABBREVIATIONS: ABBREVIATIONS: OPMD = oculopharyngeal MD; LGMD = limb-girdle MD; GERD = gastroesophageal reflux disease; CK = creatine kinase; EMG = electromyography

Geriatric Risk Factors and Late-Onset Subtypes

FACTOR/SUBTYPE	RISK SIGNAL	NOTES
Myotonic dystrophy type 2 (DM2)[4,32]	Diagnostic delay ~**14 years** (double DM1's ~7)	Leg weakness most common first symptom **(32.6%)**; pain first in **11.1%** vs 3.0% in DM1 (p<0.0001)
Age >65 with proximal weakness[23]	All DM2 patients >65 met formal **sarcopenia** criteria	**DM2 and sarcopenia clinically indistinguishable** without EMG + genetic testing
OPMD (late-onset, AD)[33,34]	Systematic screening found **10.5%** OPMD among unexplained myopathy	Median onset 5th-6th decade; widely underdiagnosed; **delay 3-6 years**[7,27]
Family history/de novo[12]	DMD/BMD X-linked; **~1/3 de novo**; DM and FSHD autosomal dominant	**Absent family history does not exclude MD**; genetic counseling for affected and at-risk kin
Chronic statin exposure[35]	**Statins can unmask/aggravate** late-onset MD	**Investigate persistent weakness** rather than reflexively switching or stopping the statin

ABBREVIATIONS: ABBREVIATIONS: DM1/DM2 = myotonic dystrophy type 1/2; OPMD = oculopharyngeal MD; AD = autosomal dominant; DMD/BMD = Duchenne/Becker MD; FSHD = facioscapulohumeral MD; EMG = electromyography

Diagnostic Thresholds and Confirmation

TEST/MARKER	DIAGNOSTIC CRITERION	NOTES
Serum CK[2,30]	Often **>10x** upper limit in dystrophinopathies	May be **normal or only mildly elevated** in FSHD, DM2, and late-onset LGMD - a critical geriatric pitfall
Genetic testing[30]	Identification of the causative mutation = **gold standard**	**First-line for distinctive phenotypes** (DM, FSHD, OPMD); NGS panel/WES for undifferentiated cases
EMG[30]	**Myotonic discharges support myotonic dystrophy**	Limited value for specific subtype in non-myotonic MD
Muscle biopsy + immunohistochemistry[21]	Protein expression (dystrophin, sarcoglycans); excludes mimics	Distinguishes MD from **inflammatory myopathy**
Muscle MRI[36]	Disease-specific **fatty-infiltration** patterns	Increasingly used to guide differential and biopsy site

ABBREVIATIONS: ABBREVIATIONS: CK = creatine kinase; EMG = electromyography; NGS = next-generation sequencing; WES = whole-exome sequencing; MRI = magnetic resonance imaging; MD = muscular dystrophy

Clues to Dig Deeper

FINDING	CONTEXT -> ACTION
Normal CK in a weak older adult	**Does not rule out MD** -> if weakness is progressive/asymmetric, proceed to EMG and genetic testing anyway[30]
Isolated dysphagia or ptosis after age 45	**Consider OPMD before defaulting to stroke/GERD/MG** -> swallow eval + PABPN1 testing[20]
Cardiac conduction disease with mild limb weakness	**Laminopathy/DM may present cardiac-first** -> ECG/echo plus neuromuscular referral, not cardiology alone[37]
Weakness unimproved after physical therapy	**Reconsider the sarcopenia/OA label** -> CK, EMG, and neurology referral[23]
Any patient facing surgery with undiagnosed proximal weakness + myotonia	**Rule out myotonic dystrophy first** -> undiagnosed DM carries lethal anesthetic risk[38]

FINDING	CONTEXT -> ACTION

ABBREVIATIONS: ABBREVIATIONS: CK = creatine kinase; EMG = electromyography; OPMD = oculopharyngeal MD; GERD = gastroesophageal reflux; MG = myasthenia gravis; OA = osteoarthritis; DM = myotonic dystrophy

Common Oversights

OVERSIGHT/SHORTCUT	WHY IT MATTERS – WHAT TO DO
"It's just aging/sarcopenia"	Cloaks active dystrophic wasting -> **if decline is progressive or asymmetric, order CK and refer**[23]
Normal CK ends the workup	Falsely reassuring in DM2/FSHD/late LGMD -> **pursue EMG and genetic testing when suspicion persists**[30]
Attributing all weakness to OA/spinal stenosis	True myopathy overlaps joint disease -> **examine for proximal, symmetric-vs-asymmetric weakness pattern**[3]
Stopping/switching a statin and moving on	May be unmasking MD -> **document CK, reassess after washout**, refer if weakness persists[39]
Focusing only on mobility	Cardiac and respiratory failure can be **silent** even while **ambulatory** -> baseline ECG/echo and PFTs[10]
Trusting an ICD-coded MD in claims data without chart review	Over- and undercoding are both common in administrative data; a DMD claims algorithm reached PPV **91.6%** only with supplementary clinical markers[40,41]

ABBREVIATIONS: ABBREVIATIONS: CK = creatine kinase; EMG = electromyography; OA = osteoarthritis; DM2 = myotonic dystrophy type 2; LGMD = limb-girdle MD; PFT = pulmonary function test

Key Differentials in Elderly

PRESENTATION	DIFFERENTIAL TO EXCLUDE	KEY TEST
Proximal weakness, difficulty rising	Sarcopenia/age-related deconditioning[23]	**CK, EMG, genetic panel**
Proximal weakness + pain[26]	Osteoarthritis, spinal stenosis, radiculopathy	**Exam pattern; CK; imaging as indicated**

PRESENTATION	DIFFERENTIAL TO EXCLUDE	KEY TEST
Dysphagia +/- ptosis[27]	Stroke, GERD, esophageal dysmotility, myasthenia gravis	**Swallow eval; PABPN1 (OPMD); AChR antibody**
Myalgia on statin[42]	Statin-associated muscle symptoms	**CK; washout trial; neuromuscular referral**
Subacute symmetric weakness[43]	Inflammatory myopathy (polymyositis)	**Biopsy + immunohistochemistry to avoid unnecessary immunosuppression**

ABBREVIATIONS: ABBREVIATIONS: CK = creatine kinase; EMG = electromyography; GERD = gastroesophageal reflux; OPMD = oculopharyngeal MD; AChR = acetylcholine receptor

Comorbidity Screening

CONDITION	PREVALENCE/SIGNAL	SCREENING APPROACH
Dilated cardiomyopathy[44]	**>33%** of sarcoglycanopathy, up to **50%** of LGMD2I	Annual echo
Conduction disease/ arrhythmia[18]	AF, flutter, AV block; major mortality driver in DM1	Annual ECG, Holter if symptomatic
Chronic respiratory failure[9]	**Leading cause of death in DM1** (median age 54)	Upright+supine spirometry
Oropharyngeal dysphagia[22]	OPMD, DM1/DM2, advanced LGMD -> **aspiration risk**	Swallow eval
Osteoporosis[30]	**Immobility + corticosteroid exposure**	DEXA
Diabetes/endocrine dysfunction[32]	**DM1/DM2 insulin resistance**, thyroid, hypogonadism	Glucose tolerance and lipid monitoring

ABBREVIATIONS: ABBREVIATIONS: LGMD = limb-girdle MD; LGMD2I = FKRP-related LGMD; AF = atrial fibrillation; AV = atrioventricular; DM1/DM2 = myotonic dystrophy type 1/2; OPMD = oculopharyngeal MD; DEXA = dual-energy x-ray absorptiometry

Validated Functional Measures and Classification

INSTRUMENT/CLASSIFICATION	WHAT IT MEASURES	USE IN PRACTICE
Muscular Dystrophy Functional Rating Scale (MDFRS)[45,46]	Activity and participation across MD subtypes	**Highest-rated instrument per COSMIN systematic review**; validated across multiple MD types
Vignos (lower-extremity) + Brooke (upper-extremity) scales[47]	Ambulatory vs non-ambulatory functional stage	**Define disease stages** (early/late ambulatory, non-ambulatory); widely used in natural-history studies
North Star Ambulatory Assessment (NSAA)/6-Minute Walk Test[48,49]	Motor function; walk distance	**Validated chiefly in ambulatory DMD**; limited validation in adult/ geriatric MD
Motor Function Measure (MFM)[45]	Standing/transfers, axial/proximal, distal function	**General neuromuscular instrument**; second most recommended after MDFRS

ABBREVIATIONS: ABBREVIATIONS: MDFRS = Muscular Dystrophy Functional Rating Scale; COSMIN = Consensus-based Standards for the selection of health Measurement Instruments; NSAA = North Star Ambulatory Assessment; MFM = Motor Function Measure; DMD = Duchenne MD; MD = muscular dystrophy. THIN: no staging system is validated specifically for geriatric-onset MD – most tools derive from pediatric DMD cohorts (OE-8 evidence gap); use for trajectory tracking, not age-specific staging

Muscular Dystrophy Functional Rating Scale (MDFRS)

33-item, 4-point clinician-administered scale; validated across DMD, BMD, FSHD, LGMD subtypes.[46]

DOMAIN	ITEM (N)	SCORE RANGE	WHAT IS ASSESSED	RELEVANCE IN MD
Mobility	9	**9-36**	**Ambulatory tasks**: Stair climbing; outdoor and indoor mobility; transfers from bed to chair; wheelchair manipulation; standing from sitting; sitting from lying; rolling; changing body position in bed	**Tracks the ambulatory-to-non-ambulatory transition central to DMD, LGMD, and BMD progression**; serial scores identify the point at which assistive device or wheelchair prescription is indicated

DOMAIN	ITEM (N)	SCORE RANGE	WHAT IS ASSESSED	RELEVANCE IN MD
Basic Activities of Daily Living (ADL)	6	**6-24**	**Self-care tasks**: Feeding; combing hair; brushing teeth; dressing upper/ lower body; toileting; bathing	**Directly informs home health aide needs**, caregiver burden documentation, and functional decline justification for DME prior authorization
Arm Function	7	**7-28**	**Upper extremity tasks**: Managing objects overhead; carrying objects; cleaning table; writing; turning books; picking up small objects; manipulating small objects	**Critical in FSHD and LGMD with upper extremity predominance**; tracks independence with communication and workplace tasks; informs occupational therapy referral threshold
Impairment	11	**11-44**	**Physiologic deficits**: Upper and lower limb joint contracture severity; number of contracted joints (upper and lower limbs); neck contracture severity; neck strength; trunk strength; scoliosis; orthopnea; sputum clearance; ventilator use	**Captures steroid-related sequelae in DMD** (contractures, proximal weakness); FVC tracking supports respiratory-decline trajectory and NIV initiation timing
Total	33	**33-132**	**Higher score = better function**; lower score = greater impairment	**Track total score at each visit**; declining score over **6–12 months** supports disease progression documentation and satisfies the COA functional status sub-measure

4-Point Scoring Anchor

SCORE	MEANING
1	**Unable** to perform the activity
2	Requires **assistance** (from a person or adaptive device)
3	**Independent**, but **slower** than normal speed

SCORE	MEANING
4	Performs at **normal speed** (no impairment)

RED FLAG — EMERGENCY/ACTIVATE CRISIS RESPONSE

- **Acute respiratory failure**: SpO2 <90%, acute hypercapnia, or inability to clear secretions in known MD → ED transfer
- **Hemodynamically unstable arrhythmia or syncope**: VT, complete heart block (especially LGMD1B, EDMD, DM1) → ED/cardiology
- **Aspiration pneumonia with distress**: Acute dyspnea, fever, hypoxia in a patient with known dysphagia → ED
- **Perioperative crisis in undiagnosed myotonic dystrophy**: Prolonged apnea, myotonic crisis, or cardiovascular collapse during/after anesthesia → emergency support

RED FLAG — URGENT (24-72 HOURS)

- **New cardiac conduction abnormality**: New prolonged PR, QRS widening, or new AF → urgent cardiology for device evaluation
- **Rapid respiratory decline**: FVC drop >10% from baseline or new morning headache/ daytime somnolence → urgent pulmonology
- **Acute rhabdomyolysis**: CK >10x ULN with dark urine → same-day evaluation for renal injury
- **New dysphagia with weight loss** → urgent swallowing evaluation for aspiration/ malnutrition risk
- **Progressive proximal weakness on a statin** → CK and neuromuscular referral within 1-2 weeks

AAVBC PERSPECTIVE

AAVBC encourages primary care providers to recognize that several adult-onset forms of muscular dystrophy present or accelerate in older adults and are routinely misattributed to common geriatric conditions. **Myotonic Dystrophy Type 2** is frequently cloaked by sarcopenia, statin myopathy, or fibromyalgia. **Limb-Girdle MD** presents with proximal hip and shoulder weakness that is commonly misread as osteoarthritis, spinal stenosis, or radiculopathy. **Oculopharyngeal MD** produces dysphagia and ptosis that are attributed to stroke, GERD, or myasthenia gravis. **Emery-Dreifuss MD** and late-onset LGMD may present cardiac-first (with conduction block or arrhythmia) before limb weakness emerges. The detail

that separates muscular dystrophy from normal aging is **asymmetric muscle wasting**: one limb or shoulder wasting faster than the other does not occur in typical sarcopenia and should not be attributed to it. Adult-onset diagnostic delay is prevalent and the diagnosis is generally available earlier to the provider who recognizes it. This helps ensure the right patients are identified and referred for the right evaluation at the right time.

3 MEAT DOCUMENTATION ESSENTIALS

MEAT documentation -- **Monitor, Evaluate, Assess, Treat** – is how an MD diagnosis is credibly supported and redocumented each year. Good documentation reflects the clinical reality of a systemic disease; comprehensive coding follows from it, not the reverse.

A 71-year-old man is seen for "getting weaker." Over two years he has struggled to rise from chairs and climb stairs, and his wife notes his right shoulder looks thinner than his left. He takes a statin; a prior CK was "normal" and the weakness was attributed to age. On exam there is asymmetric proximal weakness. CK is mildly elevated, EMG shows a myopathic pattern, and an NGS panel confirms a limb-girdle muscular dystrophy; baseline ECG reveals a first-degree AV block.

MONITOR: "Progressive proximal weakness x2 years; asymmetric right shoulder-girdle wasting noted by spouse. Timed chair-rise test slowed compared to prior visit. **CK 640 U/L** (mildly elevated, reference <200 U/L). Baseline **FVC 78% predicted**. ECG: **PR interval 0.24 s**, consistent with first-degree AV block."

EVALUATE: "EMG: myopathic pattern without denervation. NGS neuromuscular panel: positive for limb-girdle muscular dystrophy, sarcoglycan-associated subtype pending further classification. Echocardiogram: **LVEF 55%**, no wall-motion abnormality. Swallow screen: negative. Statin exposure reviewed — weakness pre-dates statin initiation and persisted on a supervised statin holiday; not attributed to statin myopathy."

ASSESS: "Limb-girdle muscular dystrophy, genetically confirmed (**G71.038**), with first-degree AV block (**I44.0**) and mild baseline restrictive pulmonary pattern. Not consistent with age-related sarcopenia. Systemic disease requiring ongoing cardiac and respiratory surveillance."

TREAT: "(1) Referral to multidisciplinary neuromuscular clinic. (2) Cardiology referral for conduction monitoring; annual ECG and echocardiogram. (3) Pulmonology baseline evaluation with upright and supine spirometry. (4) PT/OT referral for safe transfer technique and fall prevention. (5) Genetic counseling offered to patient and adult children. (6) Anesthetic precautions (malignant hyperthermia-adjacent risk, avoid depolarizing agents) flagged in chart for future procedures."

Documentation Elements That Protect the Record

- **Name the subtype and its genetic/CK basis** – link "weakness" to the confirmed dystrophy, not a symptom code
- **Specify the ICD-10 to the protein level** when known (calpain-3, dysferlin, sarcoglycan, anoctamin-5, FKRP) rather than defaulting to **G71.00**
- **Document organ involvement with its own code** – cardiomyopathy, conduction disease, respiratory failure, dysphagia – even while ambulatory

- **Record CK values with dates** and note explicitly that a normal CK does not exclude MD
- **Reconfirm the diagnosis at least annually** – HCC 197 must be documented and coded each year to persis

Reframing Common Documentation Shortcuts

INSTEAD OF...	DOCUMENT...
"Generalized weakness" (R53.1)	"Limb-girdle muscular dystrophy, dysferlin type **(G71.033)**, genetically confirmed"
"Muscular dystrophy" (G71.00)	The specific subtype code (e.g., **G71.02** FSHD; **G71.11** myotonic) once known
"Cardiomyopathy" listed in isolation	"Dilated cardiomyopathy **(I42.0)** secondary to muscular dystrophy" -- linked etiology
"Dysphagia"	"Oropharyngeal dysphagia (R13.12) due to OPMD; aspiration-risk plan"
"Contractures"	Joint **(M24.5-)** or muscle **(M62.4-)** contracture with **exact anatomical site**
"On home ventilator"	"Chronic respiratory failure **(J96.1-)**, ventilator dependence **(Z99.11)**"

ABBREVIATIONS: ABBREVIATIONS: FSHD = facioscapulohumeral MD; OPMD = oculopharyngeal MD; FKRP = fukutin-related protein; CK = creatine kinase; HCC = Hierarchical Condition Category

DOCUMENTATION REMINDER IS COMPREHENSIVE CODING

The most common documentation gap in muscular dystrophy is the unlinked symptom code: "generalized weakness" or an isolated "cardiomyopathy" documented without reference to the underlying dystrophy. **Each manifestation should be linked to the confirmed subtype, organ involvement should be coded separately, and the diagnosis should be reconfirmed annually.** This approach ensures the record accurately reflects the systemic nature of the disease.

For instance, a patient with confirmed Duchenne MD presenting with cardiomyopathy should be documented as **G71.01** (Duchenne muscular dystrophy) + **I43** (Cardiomyopathy in diseases classified elsewhere), not **I42.9** (Cardiomyopathy, unspecified) in isolation, which severs the link to the underlying dystrophy.

4 TREATMENT AND REFERRAL QUICK GUIDE

Management is anchored in the **DMD Care Considerations**, the **AAN/AANEM LGMD guideline** (2014), and the **AHA statement on cardiac involvement in neuromuscular disease** (2017); for adult-onset subtypes, care is primarily supportive and multidisciplinary.[10,16,17]

Therapy Escalation Criteria

TRIGGER	ACTION*
New MD diagnosis (any subtype)[48]	**Refer to a multidisciplinary neuromuscular clinic**; establish cardiac and respiratory baselines
At-risk subtype (DMD/BMD, LGMD, sarcoglycanopathy, EDMD, DM1/DM2)[18]	ECG + echocardiography at diagnosis and **annually**; Holter if conduction abnormality or symptoms
FVC <80% predicted with respiratory symptoms, or FVC ≤50% predicted regardless of symptoms, or nocturnal symptoms (orthopnea, morning headache, unrefreshing sleep)[50]	**Urgent pulmonology**; assess for noninvasive ventilation (NIV)
Symptomatic myotonia (DM1/DM2)[51]	**Trial mexiletine, flecainide, or propafenone** - expert-consensus level; see medication-safety cautions
Confirmed DMD (pediatric-derived evidence)[52]	**Corticosteroid** (deflazacort or prednisone); genotype-specific therapy where amenable

ABBREVIATIONS: ABBREVIATIONS: MD = muscular dystrophy; DMD/BMD = Duchenne/Becker MD; LGMD = limb-girdle MD; EDMD = Emery-Dreifuss MD; DM1/DM2 = myotonic dystrophy 1/2; FVC = forced vital capacity; NIV = noninvasive ventilation
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Guideline-Aligned Management by Category

CATEGORY	RECOMMENDED AGENT/ APPROACH*	NOTES
Disease-modifying (DMD only)	**Deflazacort 0.9 mg/kg/day**; **prednisone/prednisolone 0.75 mg/kg/day**[53]	**Deflazacort prolonged ambulation** (mean 13.9 vs 10.0 yrs); less weight gain than prednisone[48]

CATEGORY	RECOMMENDED AGENT/ APPROACH*	NOTES
Dissociative steroid (DMD)	**Vamorolone 6 mg/kg/day once daily**[54]	FDA-approved for **DMD ≥2 yrs**; potentially fewer steroid side effects
Genotype-specific (DMD only)[55,56]	**Exon-skipping oligonucleotides**; micro-dystrophin gene therapy (delandistrogene moxeparvovec)	Accelerated approval; **~30% of DMD amenable**; benefit not definitively established per systematic review55
Supportive (all subtypes)	**PT/OT every 4-6 mo**; low-impact aerobic exercise; bracing; NIV for respiratory insufficiency[17,48]	**Avoid supramaximal exercise**; NIV improves quality of life
Myotonia symptom control (DM1/DM2)	**Mexiletine, flecainide, or propafenone**[51]	Expert consensus; see **HIGH-risk** medication cautions below

ABBREVIATIONS: ABBREVIATIONS: DMD = Duchenne MD; PT/OT = physical/occupational therapy; NIV = noninvasive ventilation; FDA = US Food and Drug Administration; DM1/DM2 = myotonic dystrophy ½
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

CLINICAL PEARL: MATCH THE SUBTYPE AND ORGAN SYSTEM

Intervention selection should be guided by the **specific muscular dystrophy subtype and the affected organ system**, rather than by the general label "muscular dystrophy." Disease-modifying drugs are specific to Duchenne muscular dystrophy and derived from pediatric populations. For adult patients with DM2, LGMD, FSHD, or OPMD, the evidence-based approach consists of surveillance (cardiac, respiratory, swallow), fall prevention, and coordinated specialist care.

Non-Pharmacologic Care and Supportive Interventions

INTERVENTION	TARGET/RECOMMENDATION	NOTES
Physical/occupational therapy	**Reassess every 4-6 months**[48]	Maintain range of motion; prevent contractures; adapt assistive devices[17]
Low-impact aerobic exercise[17]	Swimming, stationary cycling	**Safe in moderation**; avoid supramaximal/eccentric overexertion

INTERVENTION	TARGET/RECOMMENDATION	NOTES
Fall prevention, durable medical equipment and safe transfers[48]	Home assessment; braces, walker, or powered chair as frailty rises	Central to geriatric MD safety per PCP-as-coordinator role
Bone health[57]	DEXA; vitamin D/calcium; weight-bearing as tolerated	Immobility + steroid exposure raise **fracture risk**
Genetic counseling[55]	Offer to patient and at-risk adult children	Especially for autosomal dominant DM/FSHD and X-linked DMD/BMD

ABBREVIATIONS: ABBREVIATIONS: DEXA = dual-energy x-ray absorptiometry; MD = muscular dystrophy; PCP = primary care provider; DM = myotonic dystrophy; FSHD = facioscapulohumeral MD

Medication Safety and Key Interactions

DRUG CLASS	RISK LEVEL - INTERACTION	CLINICAL ACTION
Anesthetics, sedatives, neuromuscular blockers[58]	**HIGH** - undiagnosed myotonic dystrophy carries **lethal perioperative risk**: prolonged apnea, myotonic crisis (succinylcholine), arrhythmia	**Screen for DM before elective surgery**; flag anesthetic precautions prominently in the chart
Mexiletine (and antimyotonia agents)[59]	**HIGH** - mexiletine is **absolutely contraindicated** in DM1 patients with cardiac involvement	In DM1 with conduction disease, **coordinate any antimyotonia therapy with cardiology**; document cardiac status first
Statins[28]	**MODERATE** – may unmask or aggravate subclinical late-onset MD	**Do not reflexively stop/switch**; document CK, reassess after washout, refer if weakness persists
Opioids/CNS depressants[58]	**MODERATE** – additive respiratory depression in restrictive MD	**Use lowest effective dose**; monitor ventilation, especially with nocturnal hypoventilation
Corticosteroids (chronic)[49]	**INFORMATIONAL** – glucose intolerance, osteoporosis, cataract, adrenal suppression	**Heightened geriatric vigilance**; DEXA, glucose, ophthalmology surveillance

ABBREVIATIONS: ABBREVIATIONS: DM/DM1 = myotonic dystrophy (type 1); MD = muscular dystrophy; CK = creatine kinase; CNS = central nervous system; DEXA = dual-energy x-ray absorptiometry

When to Refer

CRITERION	SPECIALIST	URGENCY
Every newly diagnosed/ suspected MD[17]	Multidisciplinary neuromuscular clinic (neurology)	Routine, but expedited if rapidly progressive
New conduction abnormality or reduced LVEF[10]	Cardiology (device evaluation, cardiomyopathy therapy)	**Urgent** (same/next-day if unstable)
FVC <50% predicted as indication for NIV[50]	Pulmonology/sleep medicine (NIV)	**Urgent**
New dysphagia with weight loss[17]	Speech-language pathology; consider GI	**Urgent** (aspiration/ malnutrition risk)
Scoliosis or contracture progression[17]	Orthopedic surgery; rehabilitation	Within 1-2 weeks
Any patient + affected family[55]	Genetic counseling	Routine

ABBREVIATIONS: ABBREVIATIONS: MD = muscular dystrophy; LVEF = left ventricular ejection fraction; FVC = forced vital capacity; NIV = noninvasive ventilation; GI = gastroenterology

Follow-Up Timing

DOMAIN/STAGE	FREQUENCY	WHAT TO MONITOR
Cardiac (at-risk subtypes)	**Annually** (more often if abnormal)[10]	ECG + echocardiography; Holter if symptomatic
Respiratory	**At diagnosis, then periodically**[17]	Upright + supine spirometry; nocturnal symptoms; SpO^2
Functional status	**Every 4-6 months** (PT/OT)[48]	Chair-rise, gait, timed function; validated scale where applicable
Swallow/nutrition	**When symptomatic; then periodically**[17]	Dysphagia screen; weight; aspiration risk

ABBREVIATIONS: ABBREVIATIONS: PT/OT = physical/occupational therapy; HCC = Hierarchical Condition Category; SpO2 = peripheral oxygen saturation

Comorbidity Management

COMORBIDITY	PRIMARY CARE APPROACH	CAUTION
Cardiomyopathy/conduction disease[10]	**Coordinate ACEi/beta-blocker and device decisions** with cardiology	**May be silent while ambulatory** – surveillance, not symptom-driven
Restrictive respiratory failure[50]	**Early NIV referral**; vaccinate; aggressive infection management	**Patients often lack dyspnea before failure** – PFTs are the detector
Dysphagia/malnutrition[17]	**Swallow-safe strategies**; dietitian	**Aspiration pneumonia risk**; monitor weight
Diabetes/endocrine (DM1/DM2)[60]	**Glucose tolerance and lipid monitoring**; treat per standard targets	**Insulin resistance, thyroid dysfunction, hypogonadism** common
Osteoporosis[61]	**DEXA**; vitamin D/calcium; treat fracture risk	Compounded by **immobility and steroid exposure**

ABBREVIATIONS: ABBREVIATIONS: ACEi = ACE inhibitor; NIV = noninvasive ventilation; PFT = pulmonary function test; DM1/DM2 = myotonic dystrophy 1/2; DEXA = dual-energy x-ray absorptiometry

Patient Education and Adherence

- **Systemic disease framing**: Patients and families should understand that MD is systemic, not just "weak muscles"
- **Warning signs requiring immediate action**: Teach recognition of breathing difficulty or trouble clearing secretions, palpitations or fainting, and choking or new swallowing trouble
- **Appointment adherence**: Emphasize the importance of keeping cardiac and pulmonary appointments even when feeling well
- **Genetic counseling**: Highlight its value for adult children
- **Advance-care and transitional planning**: Part of ongoing care
- **Emotional wellbeing checks**: Part of ongoing care

DOCUMENTATION REMINDER - PATIENT EDUCATION

Document the education provided and the patient's functional trajectory; assistive-device needs, fall history, and advance-care discussions. This ensures the record reflects the real burden of a progressive systemic disease rather than a single static diagnosis.

Cost-Smart Options

The following cost comparisons apply specifically to **Duchenne muscular dystrophy (DMD)**, which has the largest portfolio of FDA-approved therapies among the muscular dystrophy subtypes. Other forms — including **myotonic dystrophy**, **limb-girdle muscular dystrophy**, and **facioscapulohumeral muscular dystrophy** — currently have **no FDA-approved disease-modifying therapies**; management in those subtypes is supportive and symptom-directed. Confirm subtype documentation before applying any treatment or cost guidance below.

BRAND (EST. COST)*	GENERIC/ ALTERNATIVE (EST. COST)*	EST. SAVINGS	COST-SMART TIP
Brand Emflaza (deflazacort) 6mg oral tablet (~$10,003/100 tablets; ~$9,003/month at standard 3-tablet/day dosing)[62]	Generic **deflazacort** 6mg (**~$6,884/100 tablets**; ~**$6,195/month**)[63] — or generic **prednisone** (~**$9–15/month**) as dramatically lower-cost corticosteroid alternative[64]	Generic deflazacort: **~$5000/ month** vs. brand Emflaza; generic prednisone: **~$9000/ month** vs. brand Emflaza	Generic deflazacort is available and appropriate — prescribe generic over brand Emflaza with no clinical justification for brand; generic **prednisone** is dramatically cheaper but is **not FDA-approved for DMD** and has shown inferior muscle preservation vs. deflazacort in head-to-head data — corticosteroid selection should be confirmed with neuromuscular specialist
Brand Exondys 51 (eteplirsen IV weekly) (~$1,571/100mg vial)[65] **— at standard dosing of 30 mg/kg/week for avg 70 kg adult = ~21 vials = ~$131,944/month**	**No generic exists** — mutation-specific therapy indicated only for confirmed **exon 51 skipping-amenable DMD mutations**; no lower-cost alternative within this class	N/A	Exondys 51 is specialist-prescribed and managed — PCP role is to document **confirmed exon 51-amenable mutation** on genetic testing to support prior authorization; billed under **Medicare Part B** (IV-administered), not Part D — **IRA Part D $2,100 OOP cap does not apply**; all payers require prior authorization

BRAND (EST. COST)*	GENERIC/ ALTERNATIVE (EST. COST)*	EST. SAVINGS	COST-SMART TIP
Brand Elevidys (delandistrogene moxeparvovec-rokl), one-time IV gene therapy, WAC (~$3.2 million per infusion)[66,67]	**No alternative exists** — first and only FDA-approved gene therapy for DMD; single lifetime administration	**N/A**	Elevidys is among the most expensive medicines in the US; most patients do not pay full WAC — Sarepta outcomes-based payer agreements and net price reductions of **~20%** are in place; **all payers require prior authorization** and most require administration at a designated **Gene Therapy Center of Excellence**; PCP documentation of **confirmed DMD gene mutation**, current **ambulatory status**, and **patient weight** is required to initiate authorization; enroll eligible patients in Sarepta's **myDuchenne** support program proactively

ABBREVIATIONS: DMD = Duchenne muscular dystrophy; IV = intravenous; WAC = wholesale acquisition cost; IRA = Inflation Reduction Act; OOP = out-of-pocket; MPPP = Medicare Prescription Payment Plan; Part B/D = Medicare Parts B and D ***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Quality Metrics Tie-In

Muscular dystrophy has no dedicated HEDIS measure or CMS Star Rating. The measures below reflect adjacent clinical priorities, such as fall prevention, bone health, cardiometabolic comorbidities, and functional status. These are broadly applicable to this population within primary care and value-based care settings.

MEASURE[68]	STANDARD	APPLICABILITY TO MD
Care for Older Adults (COA)	**Denominator:** MA enrollees ≥66, continuously enrolled **Numerator:** 2 sub-measures documented annually: functional status assessment, medication review **Exclusions:** hospice, ESRD	**COA is the most broadly applicable cross-cutting measure** given progressive functional decline, polypharmacy (corticosteroids, antiarrhythmics, ACE inhibitors), and the high burden of advance care planning in this population
Falls: Screening for Future Fall Risk (MIPS #318)	**Denominator:** patients ≥65 with a qualifying encounter during the performance period **Numerator:** screened for future fall risk during the measurement period; **Exclusions:** hospice	**Age-gated to ≥65**; applies to older adults with limb-girdle, facioscapulohumeral, or late-presenting MD. For younger MD patients, use MIPS #513 instead
Falls: Plan of Care (MIPS #155)	**Denominator:** patients ≥65 with history of falls (2+ falls or any fall with injury in the past year) and a qualifying encounter **Numerator:** plan of care documented including balance, strength, and gait training **Exclusions:** hospice	**Triggered by documented fall history**; age-gated to ≥65. Document plan at the visit where fall history is identified to satisfy the measure at that encounter
Patient Reported Falls and Plan of Care (MIPS #513)	**Denominator (any age):** patients with an active diagnosis of a movement disorder, neuromuscular disorder, multiple sclerosis, dementia, or stroke **Numerator (Strata 2, scored):** plan of care for falls documented for patients who reported a fall since last visit **Exceptions:** documented medical reason for not assessing (e.g., syncope, epilepsy, concussion/mTBI)	**The most directly applicable falls measure for MD** — "neuromuscular disorder" is explicitly in the denominator with no age restriction. Documents Duchenne, Becker, and limb-girdle MD patients of any age. Strata 1 (fall incidence tracking) is data-completeness only and not scored

MEASURE[68]	STANDARD	APPLICABILITY TO MD
Osteoporosis Screening (MIPS #39)	**Denominator:** women 65-85 without a prior osteoporosis diagnosis, with a qualifying encounter **Numerator:**documentation of central DXA of hip or spine, ever performed **Exclusions:** prior diagnosis of osteoporosis; hospice	**Applies to women 65-85 without an existing osteoporosis diagnosis.** For MD patients with immobility-related bone loss who are younger, male, or already diagnosed — proactive DEXA at the start of chronic corticosteroid therapy is an internal benchmark with no external measure equivalent
Osteoporosis Management after Fracture (HEDIS OMW; MIPS #418)	**Denominator:** women 50-85 with a fracture during the measurement period or year prior **Numerator:** BMD test or pharmacotherapy for osteoporosis prescribed within 180 days of fracture **Exclusions:** fractures of finger, toe, face, or skull; hospice; frailty + advanced illness (≥66)	**Post-fracture only; applies to women 50-85.** Steroid-induced bone loss in Duchenne MD increases fracture risk substantially — document fracture events explicitly to trigger this measure and ensure pharmacologic treatment is initiated within the 180-day window
Diabetes: Glycemic Status Assessment (HEDIS GSD; MIPS #1)	**Denominator:** patients 18-75 with diabetes (≥2 diagnoses on different service dates in measurement period or prior year, or diagnosis + dispensed diabetes medication) **Numerator (inverse):** most recent HbA1c > 9.0% or missing **Exclusions:** hospice, palliative care, frailty + advanced illness (≥66)	**MIPS #1 is an inverse measure.** Myotonic dystrophy type 1 and 2 cause insulin resistance via impaired insulin receptor splicing; code secondary diabetes as E08.– rather than E11.– when related to the underlying neuromuscular disorder. Corticosteroid use in Duchenne MD additionally elevates glucose independent of myotonic dystrophy mechanisms

MEASURE[68]	STANDARD	APPLICABILITY TO MD
Controlling High Blood Pressure (HEDIS CBP; MIPS #236)	**Denominator:** patients 18-85 with hypertension (≥2 outpatient visits with HTN diagnosis in measurement period or year prior) **Numerator:** most recent BP <140/90 mmHg during measurement period **Exclusions:** ESRD, hospice, palliative care, pregnancy	**Hypertension and cardiomyopathy are common comorbidities across MD subtypes**; corticosteroid use in Duchenne MD additionally elevates BP. Cardiac disease is the leading cause of death in many MD subtypes — BP control is a direct cardiac risk-reduction lever in this population

ABBREVIATIONS: ACE = angiotensin-converting enzyme; ACP = advance care planning; BMD = bone mineral density; BP = blood pressure; CBP = Controlling High Blood Pressure; COA = Care for Older Adults; DEXA = dual-energy X-ray absorptiometry; DM1/DM2 = myotonic dystrophy type 1/type 2; ESRD = end-stage renal disease; GSD = Glycemic Status Assessment; HbA1c = hemoglobin A1c; HEDIS = Healthcare Effectiveness Data and Information Set; MA = Medicare Advantage; MD = muscular dystrophy; MIPS = Merit-based Incentive Payment System; mTBI = mild traumatic brain injury; OMW = Osteoporosis Management in Women Who Had a Fracture

QUALITY OUTCOME

There is no HEDIS or Star measure specific to muscular dystrophy. The quality that matters is reliable recognition and coordinated surveillance: catching cardiac and respiratory decline early and preventing falls and aspiration.

AAVBC PERSPECTIVE

AAVBC encourages primary care providers to abandon the assumption that adult-onset or progressive motor weakness in elderly patients represents normal aging, and to initiate a targeted neuromuscular workup when clinical features warrant it. When a geriatric patient presents with unexpected functional decline, frequent falls, progressive **proximal weakness**, or a **waddling gait**, the appropriate first step is a **serum Creatine Kinase (CK)** test. An elevated CK indicates muscle fiber breakdown and is inconsistent with typical age-related sarcopenia. Weakness that fails to improve with physical therapy is a further signal that warrants immediate referral for **EMG or targeted genetic panel sequencing** rather than continued conservative management.

AAVBC supports timely neuromuscular evaluation as an active clinical obligation in this population, recognizing that delayed diagnosis compounds functional decline, missed cardiac surveillance, and avoidable loss of independence in patients whose condition was identifiable earlier. This helps ensure the **right patients receive the right workup and the right referral at the right time**.

5 CODING REMINDERS AND CASE EXAMPLES

Coding Specificity

SPECIFICITY DISTINCTION	WHY IT MATTERS FOR CODING
Subtype vs. unspecified	Assign the named-subtype code once known; reserve **G71.00** for genuine pending-workup only
Protein-level LGMD granularity	Use calpain-3 **(G71.032)**, dysferlin **(G71.033)**, sarcoglycan **(G71.0340-.0349)**, anoctamin-5 **(G71.035)**, FKRP **(G71.036)** when genetics identify the subtype
Myotonic MD placement	**G71.11** maps to HCC 197; other myotonic disorders **(G71.12-G71.19)** do NOT. Document "myotonic dystrophy" explicitly
Codes without a dedicated entry	Emery-Dreifuss, OPMD, and congenital MD are all **G71.09** (other specified MD). Name the specific dystrophy to support it
Systemic manifestations	Code cardiomyopathy **(I42.0)**, conduction disease **(I45.-)**, respiratory failure **(J96.1-)**, dysphagia **(R13.12/.13)**, and contractures **(M24.5-/M62.4-)** separately, with anatomical site
Active vs. history	MD is a lifelong active condition – code it actively each year; do not let it drift to a history/Z-code framing

ABBREVIATIONS: ABBREVIATIONS: LGMD = limb-girdle MD; FKRP = fukutin-related protein; OPMD = oculopharyngeal MD; HCC = Hierarchical Condition Category; MD = muscular dystrophy

Annual Clinical Review and Confirmation

- **Annual review requirement.** HCC 197 does not carry over automatically - muscular dystrophy must be documented and coded during a face-to-face encounter at least once each calendar year to persist in risk adjustment
- **Qualifying encounter.** In-person visits and synchronous audio-video telehealth qualify; telephone-only contact is generally insufficient to support the annual diagnosis where clinical assessment is required

- **Reconfirm the subtype, not just "MD."** Restate the specific G71 subtype and its genetic/CK basis at the annual review, and re-document active organ involvement (cardiac, respiratory, swallowing) with its own code so the systemic picture is preserved
- **Update functional status.** Record ambulatory status, assistive-device use, and any validated functional measure at least annually – this supports both continuity of the record and appropriate clinical complexity

Good Documentation – EHR Tips

TAG	SMARTPHRASE/TIP
SMARTPHRASE	.MD_VISIT – populates subtype + **G71** code, genetic/CK basis, functional status, cardiac/respiratory surveillance status, and annual-confirmation attestation
SMARTPHRASE	.MD_MEAT – prompts all four MEAT elements (Monitor weakness/CK trajectory; Evaluate EMG/genetics/echo/PFT; Assess subtype + manifestations; Treat referrals/therapy)
SMARTPHRASE	.MD_ESCALATE – documents escalation trigger (FVC drop, new conduction abnormality), new therapy or referral, guideline alignment, and specialist status
ANNUAL DX	Standing reminder to reconfirm and re-code HCC 197 with the specific subtype at the annual wellness or chronic-care visit
ALERT	Best-practice prompt: undiagnosed proximal weakness + myotonia -> screen for myotonic dystrophy before any elective surgery (anesthetic-risk flag)

ABBREVIATIONS: ABBREVIATIONS: MEAT = Monitor/Evaluate/Assess/Treat; CK = creatine kinase; EMG = electromyography; PFT = pulmonary function test; FVC = forced vital capacity; HCC = Hierarchical Condition Category

Brief Case Examples

SUCCESS – Systemic Disease Recognized, Specific Coding, Coordinated Care

Patient: 72-year-old woman with 3 years of progressive difficulty rising from chairs; prior CK "normal," weakness attributed to age.

Documentation: PCP noted asymmetric proximal weakness, ordered CK and EMG, referred to neurology; NGS confirmed FSHD **(G71.02)**. Baseline ECG, echo, and spirometry documented; cardiomyopathy surveillance plan recorded.

Outcome: Systemic disease recognized; annual cardiac/respiratory surveillance in place; falls-prevention referral made. HCC 197 documented with specific subtype.

RAF impact: G71.02 -> HCC 197, RAF **0.426** x $10,402.34 = **~$4,431/yr**, supporting the true resource needs of coordinated multidisciplinary care.

PITFALL – Cloaked as Aging, Complexity Omitted

Patient: 70-year-old man on a statin with 2 years of proximal weakness and mild dysphagia.

Documentation as written: "Generalized weakness (R53.1), likely age-related/statin-related. Statin switched." No CK trend, no subtype, no organ workup.

Consequence: Systemic MD cloaked as aging; no cardiac or respiratory surveillance; dysphagia unaddressed (aspiration risk). No HCC documented – record does not reflect the patient's true complexity.

RAF impact: R53.1 carries **no HCC**; the ~**$4,431/yr** HCC 197 value (RAF 0.426) is lost along with the clinical rationale for surveillance.

Fix: Order and trend CK, pursue EMG/genetic confirmation, code the specific subtype (e.g., **G71.11** if myotonic), and add cardiac/respiratory/swallow surveillance with linked secondary codes.

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