AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Friedreich Ataxias

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Friedreich ataxia (FRDA) is the **most common inherited cerebellar ataxia** - an autosomal recessive, multisystem neurodegenerative disease caused by **biallelic pathogenic variants** in the **FXN** gene that **reduce frataxin**, a mitochondrial protein, leading to **mitochondrial dysfunction, progressive sensory and cerebellar ataxia**, and a characteristic **non-neurological burden** of cardiomyopathy, diabetes, and skeletal deformity.[1,2] Roughly **96%** of individuals are homozygous for a GAA trinucleotide repeat expansion in intron 1 of FXN; the remaining **~4%** are compound heterozygous for a GAA expansion plus a point mutation or deletion.[3] The broader category of **hereditary ataxias** spans numerous genetically distinct disorders - including the spinocerebellar ataxias (SCAs) and ataxia-telangiectasia - that share progressive incoordination but differ in inheritance, age of onset, and systemic involvement.[4]

ICD-10 Codes:

Genetically confirmed Friedreich ataxia is reported with **G11.11** (Friedreich ataxia), the most specific billable code, which includes late-onset and retained-reflex variants. **G11.10** (early-onset cerebellar ataxia, unspecified) is a temporary code for use only while genetic testing is pending and should be replaced once FRDA is confirmed. Non-Friedreich hereditary ataxias use **G11.2** (late-onset cerebellar ataxia), **G11.3** (ataxia with defective DNA repair, e.g., ataxia-telangiectasia), **G11.8** (other hereditary ataxias, including SCAs and DRPLA), and **G11.9** (hereditary ataxia, unspecified). **G11.9 is commonly overused**: once FRDA is genetically confirmed, continuing to report **G11.9** understates documented disease complexity. The frequent multisystem manifestations are coded separately to represent the full clinical picture - **I42.2** or **I42.0** (hypertrophic or dilated cardiomyopathy), **E08.-** (diabetes due to an underlying condition), **R13.12/ R13.13** (oropharyngeal/pharyngeal dysphagia), **M41.4-** (neuromuscular scoliosis), and **N31.-** (neurogenic bladder).[5]

Prevalence and Burden: Friedreich ataxia affects an estimated **1 in 200,000** people and is the most common inherited cerebellar ataxia; the carrier frequency in populations of European and South Asian ancestry is approximately **1 in 60 to 1 in 100**.[2,3] Classic onset falls before age **25**, but roughly **25%** of cases present atypically as Late-Onset FRDA (LOFA, onset 26-39 years), Very-Late-Onset FRDA (VLOFA, onset >=40 years), or FRDA with Retained Reflexes (FARR).[3] Cardiac disease occurs in **40-85%** of individuals and is the leading cause of death (~**62%** of deaths); dysphagia-related bronchopneumonia accounts for about **28%** of non-cardiac deaths.[2] Disease-causing GAA repeats range from **66 to 1,700**, with larger repeats predicting earlier onset and more severe systemic involvement; normal alleles carry **<=30** repeats.

HCC/RAF V28 Mapping

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
G11.11 (Friedreich ataxia)	**HCC 200** - Friedreich and Other Hereditary Ataxias; Huntington Disease	0.279	**Friedreich ataxia confirmed by FXN GAA repeat expansion**. Includes LOFA, VLOFA, and FARR; never recode late-onset FRDA as G11[2]

ICD-10 CODE(S)	HCC CATEGORY (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
G11.8 (Other hereditary ataxias)	**HCC 200**	0.279	**Other genetically defined hereditary ataxias** (spinocerebellar ataxias, DRPLA) not elsewhere classified; confirm subtype genetically
G11.2 (Late-onset cerebellar ataxia)/ G11.3 (Cerebellar ataxia with defective DNA repair)	**HCC 200**	0.279	Non-Friedreich late-onset hereditary ataxia **(G11.2)** and ataxia-telangiectasia/DNA-repair ataxias **(G11.3)**; use only for genetically confirmed non-FRDA etiologies
G11.9 (Hereditary ataxia, unspecified)	**HCC 200**	0.279	Hereditary ataxia, **unspecified**. Acceptable **only** while testing is pending-replace with **G11.11** or the confirmed subtype once results return

ABBREVIATIONS: CNA = Community Non-Dual Eligible, Aged; RAF = Risk Adjustment Factor; HCC = Hierarchical Condition Category; FRDA = Friedreich ataxia; LOFA = Late-Onset FRDA; VLOFA = Very-Late-Onset FRDA; FARR = FRDA with Retained Reflexes; DRPLA = dentatorubral-pallidoluysian atrophy. *Annual value = CNA coefficient x ~$10,402 representative base rate, illustrative only; actual plan payment varies by county and patient segment
***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[6,7]

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

Friedreich Ataxias
~$2,902

HCC 200 · RAF 0.279

2 RECOGNITION AND DIAGNOSIS

Medicare Diagnostic Workup and Surveillance (Adult and Geriatric)

SERVICE/ SURVEILLANCE	FREQUENCY	CLINICAL RATIONALE	MEDICARE COVERAGE NOTE
FXN GAA repeat testing (TP-PCR)[2,8]	Once, at diagnosis	**Targeted Triplet Repeat-Primed PCR** detects the GAA expansion that standard NGS panels miss; reflex to full FXN sequencing if a single expanded allele is found	**Covered when medically necessary for diagnosis** (CPT 81401 or lab-specific); MA prior authorization may apply - confirm with the lab and MAC
12-lead ECG[2,9]	At least annually; sooner if symptomatic	**Cardiomyopathy is the leading cause of death**; conduction changes and arrhythmia may precede symptoms	**Covered annually** or more often with documented medical necessity (CPT 93000)
Transthoracic echocardiogram[3,9]	At least annually	Tracks progression from hypertrophic to dilated phenotype and falling LVEF(AHA Class I, Level of Evidence C)	**Covered annually** for documented cardiomyopathy surveillance (CPT 93306)
Fasting glucose and HbA1c[2,10]	Annually; OGTT if impaired	**Diabetes** prominence and **carbohydrate intolerance**; insulin deficiency develops insidiously	**Covered for diabetes** screening and monitoring (CPT 83036/82947)
Swallowing evaluation (clinical or FEES)[11]	At diagnosis and with any decline	Dysphagia is present in patients and drives aspiration risk; phase-specific assessment guides diet and SLP referral	**Covered when dysphagia** is documented (CPT 92610/92612)

SERVICE/ SURVEILLANCE	FREQUENCY	CLINICAL RATIONALE	MEDICARE COVERAGE NOTE
PT/OT evaluation[3]	At diagnosis and periodically	Establishes a functional baseline and an individualized program shown to be safe and feasible in FRDA	**Covered under Part B with medical-necessity documentation** (CPT 97163/97165-97167)

ABBREVIATIONS: TP-PCR = Triplet Repeat-Primed PCR; NGS = next-generation sequencing; FXN = frataxin gene; LVEF = left ventricular ejection fraction; AHA = American Heart Association; OGTT = oral glucose tolerance test; FEES = fiberoptic endoscopic evaluation of swallowing; SLP = speech-language pathology; PT = physical therapy; OT = occupational therapy; MA = Medicare Advantage; MAC = Medicare Administrative Contractor; CPT = Current Procedural Terminology

Subtle Signs in Older Adults

SUBTLE SIGN	WHY IT IS EASILY MISSED	WHAT TO LOOK FOR
Late-onset progressive gait ataxia[2]	**Attributed to age, osteoarthritis, or deconditioning** rather than a hereditary disease	**Progressive wide-based unsteadiness** with impaired tandem gait that outpaces musculoskeletal findings; **ask about falls**
Lost vibration and proprioception[2]	**Confused with diabetic or age-related peripheral neuropathy**	**Reduced vibration and joint-position sense** at the toes with a positive Romberg, often with absent ankle reflexes
Retained or brisk reflexes with extensor plantars[2]	FARR and late-onset forms keep reflexes, so **FRDA is dismissed when reflexes are present**	**Spasticity with preserved reflexes** and upgoing toes; **do not exclude FRDA on intact reflexes alone**
Dysarthria and early swallowing change[3]	**Slurred speech and throat-clearing** are read as normal aging	**Scanning or slurred speech**, wet voice after liquids, and unintended weight loss
Exertional dyspnea or new edema[2]	**Cardiac involvement is overlooked** when the focus is on the neurological diagnosis	**New dyspnea, orthopnea, or peripheral edema** in a patient with known or suspected ataxia

ABBREVIATIONS: FARR = FRDA with Retained Reflexes; FRDA = Friedreich ataxia

Geriatric Risk Factors

RISK/SEVERITY MODIFIER	EFFECT	CLINICAL SIGNAL
Larger GAA1 (shorter-allele) repeat size[2,12]	Earlier onset, faster neurological progression, and higher cardiomyopathy and diabetes prevalence	**GAA1 length** accounts for **36-50%** of the variability in age of onset; report repeat sizes when available
Earlier age of onset[2]	More severe trajectory and earlier loss of ambulation	Onset **before age 8** progresses roughly **twice as fast** as onset after **age 15**
Cardiomyopathy[2,13]	Leading cause of mortality **(~62% of deaths)**	Lower LVEF and higher LV mass index predict mortality; the Weidemann FA-CM stage tracks severity
Diabetes mellitus[10]	Confers a prognostic disadvantage and complicates management	Reported in **~8.7-15%**; carbohydrate intolerance in **~25%**; insulin deficiency with peripheral resistance
European/South Asian ancestry[3]	Higher carrier frequency	Carrier rate **1 in 60 to 1 in 100**; relevant to family counseling and sibling testing

ABBREVIATIONS: GAA1 = shorter (driver) GAA repeat allele; LVEF = left ventricular ejection fraction; LV = left ventricular; FA-CM = Friedreich ataxia cardiomyopathy classification

Diagnostic Thresholds (FXN Genetic Confirmation and Validated Scales)

INSTRUMENT	WHAT IT MEASURES	SCORING/THRESHOLD	NOTES
FXN GAA repeat analysis (TP-PCR)	Genetic confirmation of FRDA	Biallelic GAA expansion; disease-causing repeats **66-1,700** (repeats <100 rarely cause disease; normal <=30)[2,3]	**Diagnostic gold standard**; NGS panels miss the expansion. Reflex to full sequencing if only one expanded allele

INSTRUMENT	WHAT IT MEASURES	SCORING/THRESHOLD	NOTES
Modified Friedreich Ataxia Rating Scale (mFARS)[2]	Neurological function across bulbar, upper-limb, lower-limb, and stability domains	Score 0-99; **higher = worse**. Primary endpoint in the omaveloxolone registration trial[14]	**Sensitive at early stages**; recommended for FRDA by the Movement Disorder Society
Scale for the Assessment and Rating of Ataxia (SARA)	Ataxia severity across 8 items	Score 0-40; natural-history change **~0.40 points/12 months**[2]	**Faster than mFARS**; validated for FRDA and SCAs and used in the EFACTS cohort[15]
Weidemann FA-CM classification[16]	Echocardiographic cardiac staging	Four groups (No/Mild/ Intermediate/Severe CM, the last with **EF <50%**)	Validated in 205 patients; **cardiac and neurological severity progress independently**

ABBREVIATIONS: TP-PCR = Triplet Repeat-Primed PCR; FXN = frataxin gene; FRDA = Friedreich ataxia; NGS = next-generation sequencing; mFARS = modified Friedreich Ataxia Rating Scale; SARA = Scale for the Assessment and Rating of Ataxia; SCA = spinocerebellar ataxia; EFACTS = European Friedreich's Ataxia Consortium for Translational Studies; FA-CM = Friedreich ataxia cardiomyopathy; EF = ejection fraction; CM = cardiomyopathy

Clues to Dig Deeper

CLUE	WHAT IT SUGGESTS	NEXT STEP
Progressive ataxia with absent ankle reflexes and extensor plantars[10]	Classic sensory-cerebellar FRDA pattern	**Order targeted FXN GAA testing** (TP-PCR), not a broad NGS panel alone
Adult-onset ataxia with spasticity and preserved reflexes[17]	Possible LOFA, VLOFA, or FARR mimicking hereditary spastic paraplegia	**Do not exclude FRDA on age or intact reflexes**; pursue genetic confirmation
Negative standard NGS panel in a clinically suspicious patient[17]	**NGS cannot reliably detect the GAA** repeat expansion	**Specifically request TP-PCR** for the FXN GAA repeat
Unexplained hypertrophic cardiomyopathy plus gait change[2]	Cardiomyopathy may be the first system to declare itself in FRDA	**Echocardiogram and ECG**, then coordinated neurology and cardiology evaluation

CLUE	WHAT IT SUGGESTS	NEXT STEP
New diabetes that behaves atypically for type 2[2]	FRDA-related diabetes blends beta-cell failure with insulin resistance[18]	Consider an underlying-condition etiology; **coordinate insulin-first management with endocrinology**

ABBREVIATIONS: FRDA = Friedreich ataxia; FXN = frataxin gene; TP-PCR = Triplet Repeat-Primed PCR; NGS = next-generation sequencing; LOFA = Late-Onset FRDA; VLOFA = Very-Late-Onset FRDA; FARR = FRDA with Retained Reflexes; ECG = electrocardiogram

Common Oversights

COMMON OVERSIGHT	CONSEQUENCE*	BETTER PRACTICE
Excluding FRDA because onset is after age 25 or reflexes are intact[17]	Late-onset and retained-reflex variants **(~25% of cases)** are missed and mislabeled as another ataxia	**Keep FRDA on the differential for any progressive adult ataxia and confirm genetically**
Relying on a negative NGS panel to rule out FRDA[17]	The GAA repeat expansion is invisible to standard NGS, so a true case is dismissed	**Request TP-PCR** specifically when FRDA is suspected
Treating FRDA-related diabetes as ordinary type 2[2,19]	Metformin, a Complex I inhibitor, is mechanistically concerning in a mitochondrial disease	**Favor insulin** and coordinate an FRDA-appropriate regimen with endocrinology
Reflexively reading an elevated BNP as heart-failure decompensation[14]	On omaveloxolone, **14%** have drug-induced BNP elevation, prompting unnecessary escalation	Correlate BNP with clinical signs **before** activating heart-failure pathways
Overlooking insidious dysphagia until aspiration occurs[2]	Aspiration pneumonia accounts for **~28%** of non-cardiac deaths	**Screen swallowing proactively** and refer to SLP at the first wet voice or weight loss
Missing neuromuscular scoliosis, urinary, and mood symptoms[15]	Scoliosis **(~74%)** and urinary dysfunction **(~43%)** are common but under-documented	**Examine the spine**, ask about continence, and screen for depression at each visit

ABBREVIATIONS: FRDA = Friedreich ataxia; NGS = next-generation sequencing; TP-PCR = Triplet Repeat-Primed PCR; BNP = B-type natriuretic peptide; SLP = speech-language pathology
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Key Differentials in Elderly

DIFFERENTIAL	DISTINGUISHING FEATURES	HOW TO CONFIRM/ SEPARATE
Spinocerebellar ataxias (SCAs)[20]	**Often autosomal dominant with a family history**; cerebellar signs predominate	Genetic panel for SCA repeat expansions; code **G11.8** when an SCA is confirmed
Hereditary spastic paraplegia[17,21]	**Spasticity dominates with comparatively mild ataxia**	**FRDA with spasticity can mimic this**; pursue FXN testing before settling on HSP
Ataxia-telangiectasia and DNA-repair ataxias[22]	Oculocutaneous telangiectasia, immune deficiency, cancer predisposition	**Elevated AFP and immunoglobulin deficiency**; confirm with **ATM gene testing** (chromosomal breakage/ radiosensitivity studies support diagnosis)
Acquired sensory neuropathy (diabetic, B12, alcohol)[23]	**Sensory loss** without the cerebellar and cardiac pattern of FRDA	**B12, glucose, and history FRDA** shows progressive cerebellar signs and systemic involvement
Vitamin E deficiency ataxia[24]	Phenocopies FRDA but is **treatable with supplementation**	**Serum vitamin E**; code **G11.19** for confirmed non-Friedreich early-onset ataxia

ABBREVIATIONS: SCA = spinocerebellar ataxia; FRDA = Friedreich ataxia; HSP = hereditary spastic paraplegia; FXN = frataxin gene; B12 = vitamin B12

Comorbidity Screening

SYSTEM	WHAT TO SCREEN FOR	FREQUENCY/TOOL
Cardiac[2]	Hypertrophic-to-dilated cardiomyopathy, heart failure, and arrhythmia	**Annual ECG and echocardiogram**; cardiac MRI when fibrosis assessment is indicated[9]
Endocrine[10]	Diabetes mellitus and carbohydrate intolerance	**Annual HbA1c and fasting glucose**; OGTT if impaired
Swallowing/respiratory[2]	Dysphagia and aspiration risk	**Periodic swallowing evaluation**; modified barium swallow when aspiration is suspected

SYSTEM	WHAT TO SCREEN FOR	FREQUENCY/TOOL
Musculoskeletal[15]	Neuromuscular scoliosis and restrictive respiratory compromise	**Spine exam and FVC monitoring**
Genitourinary[25]	Neurogenic bladder, urinary incontinence or retention	**Continence history** at each visit; specify dysfunction type
Psychological and sensory[2]	Depression, sleep disturbance, hearing loss, and optic atrophy	**PHQ-9 at each visit**; audiology every 2-3 years and ophthalmology assessment

ABBREVIATIONS: ECG = electrocardiogram; MRI = magnetic resonance imaging; OGTT = oral glucose tolerance test; HbA1c = glycated hemoglobin; FVC = forced vital capacity; PHQ-9 = Patient Health Questionnaire-9

⚠ RED FLAG — EMERGENCY/ACTIVATE CRISIS RESPONSE

- **Acute heart-failure decompensation** — new or worsening dyspnea, orthopnea, edema, or rapid weight gain (**≥3 lb/day** or **≥5 lb/week**), especially with falling **LVEF** or a new arrhythmia — cardiac dysfunction is the **most frequent cause of death** in FRDA
- **Symptomatic arrhythmia** (sustained palpitations, syncope, or presyncope) — **supraventricular tachycardia** occurs in **~40%** of patients with cardiac involvement
- **Aspiration pneumonia** with respiratory distress (fever, hypoxia, productive cough) in a patient with known dysphagia
- **Diabetic ketoacidosis**, which does occur in FRDA-related diabetes

⚠ RED FLAG — URGENT (24-72 HOURS)

- New **BNP elevation >200 pg/mL** in a patient on **omaveloxolone** — obtain urgent cardiology input to separate drug-induced elevation from true heart failure rather than reflexively stopping the drug
- **ALT or AST >5x ULN** on omaveloxolone — discontinue the drug and repeat liver tests, with hepatology input if bilirubin is also elevated
- New or rapidly worsening **dysphagia** with weight loss or aspiration events — same-day **SLP** and consideration of a modified barium swallow
- Acute functional decline such as **sudden loss of ambulation** — prompting urgent neurology evaluation to exclude superimposed pathology (e.g., cord compression from severe scoliosis)

3 MEAT DOCUMENTATION ESSENTIALS

Because hereditary ataxia maps to a single HCC regardless of subtype, the medical record's value comes from how completely each active manifestation is monitored, evaluated, assessed, and treated (MEAT). For Friedreich ataxia, the most important documentation is the systemic burden — cardiac, endocrine, swallowing, and skeletal.

*Case vignette: A female patient, 71, with genetically confirmed Late-Onset Friedreich ataxia is seen for a routine visit. She reports new shortness of breath climbing stairs and an unintended 6-pound weight loss. The note records 'ataxia, stable' and continues her omaveloxolone. No cardiac symptoms, swallowing status, or diabetes review are documented, and the encounter is coded **G11.9**. The next month she is hospitalized with aspiration pneumonia and newly recognized heart failure - complications that her record had never documented.*

Monitor: "mFARS/SARA score: pending, last checked >12 months ago — overdue. ECG and echocardiogram: not on file since diagnosis — overdue, order today given new exertional dyspnea. HbA1c: not rechecked this year — overdue. Swallowing status: not screened — overdue. Weight: **6-lb unintentional loss** since last visit. Omaveloxolone monitoring: liver enzymes not rechecked since initiation — overdue; BNP not drawn despite new dyspnea — order today; lipid panel not on file — overdue."

Evaluate: "New exertional dyspnea and unintentional weight loss in a patient with Late-Onset Friedreich ataxia on omaveloxolone — cannot be attributed to omaveloxolone alone without a BNP and echocardiogram, since cardiac decompensation must be excluded first. Prior note ('ataxia, stable') did not document cardiac, swallowing, or endocrine status, so today's findings represent a new evaluation, not a change from a documented baseline. Swallowing has never been formally screened despite known FRDA-associated risk — needed before an aspiration event occurs, not after."

Assess: Late-Onset Friedreich ataxia (**G11.1**), genetically confirmed. New exertional dyspnea and weight loss — **rule out hypertrophic cardiomyopathy secondary to Friedreich ataxia** pending today's echocardiogram and BNP; **rule out drug-induced BNP elevation from omaveloxolone** as an alternative explanation. Diabetes screening status: HbA1c overdue, unknown current glycemic control. Swallowing status: unknown, screening ordered today. Prior documentation ('ataxia, stable') did not reflect this systemic complexity."

Treat: "Continue **omaveloxolone**; order **BNP** and **liver enzymes** today given overdue monitoring and new symptoms. Order **echocardiogram** and **ECG** today given new exertional dyspnea — hold heart-failure-directed therapy pending results. Order **HbA1c** — if diabetes is confirmed or already present, use **insulin, not metformin**, per FRDA-specific management. Refer to **SLP** for formal swallowing evaluation given unscreened status and new weight loss. Refer to **cardiology** for co-management given new cardiac symptoms in FRDA. **PT/OT** referral to reassess functional status given weight loss and new symptoms."

Clinical Documentation Elements

- **Genetic basis:** 'Friedreich ataxia confirmed by FXN GAA repeat expansion,' supporting **G11.11** rather than an unspecified code

- **Cardiac status:** Phenotype (hypertrophic vs dilated), current LVEF, functional class, and rhythm
- **Endocrine status:** Diabetes type framed as due to the underlying condition, treatment, and most recent HbA1c
- **Swallowing and weight:** Dysphagia phase and severity, aspiration risk, and any SLP involvement
- **Musculoskeletal and urinary:** Neuromuscular scoliosis with site,[7] and bladder dysfunction type
- **Functional and mood status:** Current mFARS or SARA, mobility aids described functionally, and PHQ-9

Reframing Common Documentation Shortcuts

DOCUMENTATION SHORTCUT	WHY IT FALLS SHORT	STRONGER FRAMING
Ataxia, stable	**Omits subtype and every active comorbidity**; supports only an unspecified code	Friedreich ataxia (FXN-confirmed) with nonobstructive hypertrophic cardiomyopathy and insulin-treated diabetes - each addressed below
Manage diabetes per standard type 2 protocol	**Reproduces a metformin-first algorithm** that is mechanistically concerning in FRDA	Insulin-treated diabetes due to Friedreich ataxia; metformin avoided given Complex I inhibition - co-managed with endocrinology
Elevated BNP - rule out heart failure	**Risks treating a drug effect of omaveloxolone as decompensation**	BNP elevation interpreted in the context of omaveloxolone; no clinical heart-failure signs, so no escalation - cardiology informed
Patient is wheelchair-bound	**Labels the person** rather than describing function	Patient uses a wheelchair for mobility; transfers with assistance; PT engaged for maintenance
Scoliosis	**Idiopathic vs neuromuscular** etiology is **lost**	Neuromuscular scoliosis, thoracolumbar, secondary to Friedreich ataxia **(M41.4-)**

ABBREVIATIONS: FXN = frataxin gene; FRDA = Friedreich ataxia; BNP = B-type natriuretic peptide; PT = physical therapy; NYHA = New York Heart Association

DOCUMENTATION REMINDER IS COMPREHENSIVE CODING

Documenting the subtype and each affected system (cardiac, endocrine, swallowing, skeletal, and urinary) provides an accurate clinical picture of the patient. When documentation reflects the full extent of disease, accurate coding and patient support follow as a natural consequence of clinical documentation.

4 TREATMENT AND REFERRAL QUICK GUIDE

There is no cure for Friedreich ataxia. **Omaveloxolone**, approved in **2023**, is the first and only disease-modifying therapy — it **slows but does not halt progression**. Everything else is symptomatic, multisystem management coordinated across neurology, cardiology, endocrinology, and rehabilitation. Evidence in adults over **65** is sparse, since nearly all trial data come from ages **16-40**, so recommendations for older patients are extrapolated with added caution.[26]

Therapy Escalation Criteria

TRIGGER/FINDING	ACTION*	RATIONALE
Genetically confirmed FRDA, age >=16	Offer **omaveloxolone 150 mg** once daily on an empty stomach unless contraindicated[14]	Only FDA-approved disease-modifying therapy; **2.40-point mFARS benefit** vs placebo at 48 weeks (p=0.014)[26]
New or worsening cardiac symptoms or falling LVEF[2]	Escalate to cardiology for guideline-directed heart-failure therapy	**Cardiomyopathy is the leading cause of death**; standard HF therapy applies under specialist guidance
Rising HbA1c or new hyperglycemia	Start or intensify insulin; avoid metformin; involve endocrinology	Insulin is the mainstay (**64-74%** of FRDA-diabetes); metformin inhibits Complex I[27]
Worsening dysphagia or weight loss	Refer to SLP; modify diet textures; consider modified barium swallow	**Aspiration drives ~28% of non-cardiac deaths**; proactive management reduces pneumonia[2]
ALT/AST >5x ULN on omaveloxolone[14]	Discontinue omaveloxolone and repeat LFTs; hepatology if bilirubin elevated	Label-specified hepatic safety threshold

TRIGGER/FINDING	ACTION*	RATIONALE

ABBREVIATIONS: FRDA = Friedreich ataxia; FDA = Food and Drug Administration; mFARS = modified Friedreich Ataxia Rating Scale; LVEF = left ventricular ejection fraction; HF = heart failure; HbA1c = glycated hemoglobin; SLP = speech-language pathology; ALT = alanine aminotransferase; AST = aspartate aminotransferase; ULN = upper limit of normal; LFT = liver function test ***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Guideline-Aligned Management by Domain (GeneReviews/AHA 2017)

DOMAIN	GUIDELINE-ALIGNED APPROACH*	ELDERLY/VBC NOTE
Disease-modifying therapy[3]	**Omaveloxolone 150 mg daily**, empty stomach; dose-reduce for moderate hepatic impairment (100 mg) and **avoid in Child-Pugh C**[14]	**PK is similar across ages 16-71**, but efficacy and safety data **beyond 40** are limited
Cardiac (AHA 2017, Class I)[9]	**Annual ECG and echocardiogram**; standard ACE inhibitor/ARB/beta-blocker therapy for systolic dysfunction; cardiac MRI for fibrosis when indicated	**Interpret BNP cautiously on omaveloxolone**; transplant is an option only with mild neurological involvement
Endocrine[3,27]	**Annual HbA1c and fasting glucose**; insulin-first management; SGLT2 inhibitors or GLP-1 agonists considered with endocrinology for cardiometabolic benefit	**Avoid metformin**; in elderly patients weigh renal function and heart failure
Rehabilitation[3,9]	**Individualized** PT and OT; SLP for dysarthria and dysphagia (safe and feasible in a 2026 RCT)[28]	**Prioritize falls prevention, transfers, and adaptive seating**; continue long-term
Emerging therapy[3]	**Gene therapy, protein replacement** (nomolabofusp), and **epigenetic** approaches are in early-phase trials	Discuss trial options through a **specialized ataxia center**

ABBREVIATIONS: AHA = American Heart Association; ECG = electrocardiogram; ACE = angiotensin-converting enzyme; ARB = angiotensin receptor blocker; MRI = magnetic resonance imaging; BNP = B-type natriuretic peptide; HbA1c = glycated hemoglobin; SGLT2 = sodium-glucose cotransporter-2; GLP-1 = glucagon-like peptide-1; PK = pharmacokinetics; PT = physical therapy; OT = occupational therapy; SLP = speech-language pathology; RCT = randomized controlled trial; VBC = value-based care
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Non-Pharmacologic Care and Supportive Interventions

INTERVENTION	ROLE	PRACTICAL NOTE
Physical therapy[2,3]	Preserves strength, gait, and transfer ability	Individualized programs are safe and feasible; **integrate early and continue long-term**
Occupational therapy[3]	Maintains upper-limb function and activities of daily living	**Adaptive equipment and home assessment** as disease advances
Speech-language pathology[3]	Addresses dysarthria and dysphagia	**Swallowing strategies and diet texture changes** reduce aspiration risk
Adaptive seating/wound prevention[3]	Manages neuromuscular scoliosis and pressure-injury risk	**Seating assessment and skin checks** for wheelchair users; monitor FVC for respiratory compromise
Vitamin D and bone health[3]	Reduces fracture risk from immobility	**Annual vitamin D measurement and DXA scanning**

ABBREVIATIONS: FVC = forced vital capacity; DXA = dual-energy X-ray absorptiometry

CLINICAL PEARL: HEART AND BRAIN PROGRESS INDEPENDENTLY

In Friedreich ataxia, cardiac and neurological disease progress independently: cardiac severity does not correlate with neurological severity, so patients with mild ataxia may still have significant cardiomyopathy. Cardiac surveillance should therefore continue regardless of gait severity.

Medication Safety and Key Interactions

AGENT/CLASS*	STATUS	KEY SAFETY/INTERACTION
Omaveloxolone (Skyclarys)[14]	FDA-approved (age ≥16)	Weak CYP3A4 inducer- lowers **midazolam ~45%**, **repaglinide ~35%**, **rosuvastatin ~30%**; **avoid with moderate/strong CYP3A4 inducers**; reduce dose with CYP3A4 inhibitors; monitor LFTs, BNP, and lipids (**29%** had cholesterol above ULN)

AGENT/CLASS*	STATUS	KEY SAFETY/INTERACTION
Insulin[27]	Approved (mainstay for FRDA diabetes)	**Preferred over oral agents**; titrate with endocrinology; watch for hypoglycemia in frail elderly patients
Metformin[29]	Not contraindicated on label but **cautioned in FRDA**[30]	**Inhibits mitochondrial Complex I**, mechanistically concerning in a mitochondrial disease; expert opinion advises caution
SGLT2 inhibitors/GLP-1 agonists[27]	[OFF-LABEL] for FRDA-specific use	May offer **cardiometabolic benefit**; no FRDA-specific data-use in consultation with endocrinology
ACE inhibitors/ARBs/beta-blockers[9]	Approved (per HF guidelines)	**Standard therapy for systolic dysfunction under cardiology guidance**; avoid agents that worsen outflow obstruction in hypertrophic phenotypes

ABBREVIATIONS: FDA = Food and Drug Administration; CYP3A4 = cytochrome P450 3A4; LFT = liver function test; BNP = B-type natriuretic peptide; ULN = upper limit of normal; FRDA = Friedreich ataxia; SGLT2 = sodium-glucose cotransporter-2; GLP-1 = glucagon-like peptide-1; ACE = angiotensin-converting enzyme; ARB = angiotensin receptor blocker; HF = heart failure ***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

When to Refer

REFERRAL	WHEN	PURPOSE
Neurology[3]	**At diagnosis and ongoing**	Confirms diagnosis, initiates and monitors omaveloxolone, and manages ataxia, spasticity, and neuropathic pain
Cardiology[3]	**At diagnosis** and with any cardiac change	Surveillance, heart-failure and arrhythmia management, and transplant evaluation in selected patients
Endocrinology[3]	**On diagnosis of diabetes or carbohydrate intolerance**	Insulin-based regimen designed around FRDA-specific cautions

REFERRAL	WHEN	PURPOSE
Genetics/genetic counseling[3]	**At diagnosis**	Family counseling and testing of at-risk siblings, including echocardiographic surveillance even if asymptomatic
Palliative care[3]	**Advanced disease**	Goals-of-care planning, symptom control, and caregiver support; standard VBC frailty metrics may understate the FRDA trajectory

ABBREVIATIONS: FRDA = Friedreich ataxia; VBC = value-based care

Follow-Up Timing

DOMAIN	SUGGESTED INTERVAL	WHAT TO REVIEW
Neurological function[2,3]	Every **6-12 months**	**mFARS or SARA, falls, mobility, and omaveloxolone tolerance**
Cardiac surveillance[3]	At least **annually**	**ECG and echocardiogram with LVEF**; sooner if symptomatic
Omaveloxolone liver monitoring[14]	Baseline, **monthly x3**, then periodically	**ALT, AST, bilirubin**; discontinue if ALT/AST >5x ULN
Glycemic review[3]	**Annually**; sooner if unstable	**HbA1c and fasting glucose**; insulin titration
Swallowing, mood, and continence[3]	**Each visit**	**Dysphagia screen, PHQ-9, and bladder symptoms**

ABBREVIATIONS: mFARS = modified Friedreich Ataxia Rating Scale; SARA = Scale for the Assessment and Rating of Ataxia; ECG = electrocardiogram; LVEF = left ventricular ejection fraction; ALT = alanine aminotransferase; AST = aspartate aminotransferase; ULN = upper limit of normal; HbA1c = glycated hemoglobin; PHQ-9 = Patient Health Questionnaire-9

Comorbidity Management – Primary Care Role

COMORBIDITY	PRIMARY CARE ROLE	COORDINATION POINT
Cardiomyopathy/heart failure[3]	**Recognize symptoms early**; reinforce adherence; interpret BNP in drug context	**Co-manage with cardiology**; document phenotype and LVEF

COMORBIDITY	PRIMARY CARE ROLE	COORDINATION POINT
Diabetes[3]	**Screen and monitor**; reinforce insulin use; avoid metformin	**Endocrinology** for regimen design
Dysphagia[3]	**Screen at each visit**; arrange diet modification	**SLP** for formal evaluation and strategies
Depression/sleep[3]	**PHQ-9 screening and first-line support**	**Behavioral health and social work** as needed
Hearing/vision[3]	Ask about sensorineural loss and visual change	**Audiology every 2-3 years**; ophthalmology assessment

ABBREVIATIONS: BNP = B-type natriuretic peptide; LVEF = left ventricular ejection fraction; SLP = speech-language pathology; PHQ-9 = Patient Health Questionnaire-9

Cost-Smart Options

BRAND (EST. COST)*	GENERIC (EST. COST)*	EST. SAVINGS	COST-SMART TIP
Brand Skyclarys (omaveloxolone 150mg/day), WAC (~$370,000/year; ~$30,833/month)[31,32]	**No generic exists** — Skyclarys is the only FDA-approved disease-modifying therapy for FRDA[33]	N/A	Skyclarys is a **limited distribution drug** available only through specialty pharmacy; **96% of Medicare patients pay ≤$15/month** and 97% of commercially insured patients pay $0/month through the **Biogen REACH program**; the **IRA Part D $2,100 OOP cap** applies; PCP must document **confirmed GAA repeat expansion** and **mFARS eligibility** to support prior authorization; monitor **LFTs monthly × 3 months**, then as needed; monitor **BNP and lipids** before and during treatment[32]

BRAND (EST. COST)*	GENERIC (EST. COST)*	EST. SAVINGS	COST-SMART TIP
Brand Toprol XL (metoprolol succinate ER), avg retail (~$161/month for 30 tablets)[34]	Generic **metoprolol succinate ER** (~avg retail ~$60–65/month)[35]	~$146–150/month	Generic metoprolol succinate ER is bioequivalent to Toprol XL and is the guideline-directed beta-blocker for FRDA-associated **hypertrophic cardiomyopathy** and heart failure; no clinical indication favors brand prescribing; confirm with neurology/cardiology before initiating — beta-blocker selection and timing require specialist input in FRDA cardiac disease
Brand Zestril or Prinivil (lisinopril) — brands largely discontinued	Generic **lisinopril** (**~$4–10/month**)[36,37]	N/A	Generic lisinopril is the appropriate ACE inhibitor for FRDA-associated HF with reduced ejection fraction; prescribe generic at initiation — no brand needed; dose titration should be coordinated with cardiology
Brand Lantus (insulin glargine) (~$292–350/month)[38]	Biosimilar **Semglee** or **Basaglar** (insulin glargine) (**~$98–140/month**)[39]	~$150–250/month	FRDA-associated **diabetes mellitus** is insulin-dependent in many patients due to pancreatic beta-cell involvement; insulin glargine biosimilars are FDA-approved and interchangeable with Lantus; confirm biosimilar substitution is permitted under the patient's plan; counsel on the **IRA Part D $2,100 OOP cap** for Medicare patients

BRAND (EST. COST)*	GENERIC (EST. COST)*	EST. SAVINGS	COST-SMART TIP

ABBREVIATIONS: FRDA = Friedreich ataxia; WAC = wholesale acquisition cost; LFT = liver function test; BNP = B-type natriuretic peptide; mFARS = modified Friedreich Ataxia Rating Scale; GAA = guanine-adenine-adenine; HF = heart failure; OOP = out-of-pocket; IRA = Inflation Reduction Act; ER = extended-release
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Patient Education and Adherence

- **Systemic impact**: Explain that Friedreich ataxia affects more than balance — the heart, blood sugar, swallowing, and spine all need regular attention, even when gait feels stable
- **Treatment expectations**: Frame omaveloxolone honestly as a therapy that slows progression rather than a cure, and review its required monitoring
- **Genetic counseling**: Encourage genetic counseling for relatives
- **Support resources**: Connect families with the National Ataxia Foundation
- **Advance care planning**: Introduce goals-of-care conversations early so comfort-focused options are available when needed

DOCUMENTATION REMINDER - PATIENT EDUCATION

Document the education provided : what was discussed about cardiac and swallowing surveillance, omaveloxolone monitoring, genetic counseling, and goals of care - and the patient's understanding and preferences. This record supports continuity across the care team and Care-for-Older-Adults measures.

Quality Metrics Tie-In

MEASURE	STANDARD	APPLICABILITY TO FRDA
Falls: Screening for Future Fall Risk (HEDIS/CMS #318)[40]	**Denominator:** patients ≥65 with a qualifying encounter during the performance period **Numerator:** patient screened for future fall risk during the measurement period (document 2+ falls in past year or any fall with injury in past year) **Exclusions:** hospice	**Gait ataxia makes nearly every FRDA patient high-risk for falls regardless of age**. MIPS #318 is age-gated to ≥65; for younger FRDA patients, use MIPS #513 (see below) as the applicable falls measure. Record fall risk screening and an intervention plan at the annual visit

MEASURE	STANDARD	APPLICABILITY TO FRDA
Falls: Plan of Care (MIPS #155) and Patient-Reported Falls (MIPS #513)[41,42]	**MIPS #155 — Denominator:** patients ≥65 with history of falls (2+ falls in past year or any fall with injury in past year) and a qualifying encounter **Numerator:** plan of care documented including balance, strength, and gait training **Exclusions:** hospice **MIPS #513 — Denominator (Strata 1 and 2):** patients of any age with an active diagnosis of a movement disorder, multiple sclerosis, neuromuscular disorder, dementia, or stroke **Numerator (Strata 1, data completeness only):** patient screened for falls at each visit. **Numerator (Strata 2, performance):** plan of care for falls documented for patients who reported a fall since last visit **Exceptions:** documented medical reason for not assessing falls (e.g., syncope, vertigo, epilepsy, concussion/mTBI)	**MIPS #513 is the most directly applicable falls measure for FRDA** — "neuromuscular disorder" is explicitly in the denominator and carries no age restriction. Strata 1 of #513 (data completeness only, not scored) tracks fall incidence at every visit; Strata 2 (scored) triggers a plan of care when a fall is reported. Document a falls plan of care within 12 months for all FRDA patients regardless of age
Comprehensive Diabetes Care/ Glycemic Status (HEDIS CDC; MIPS #1)[43]	**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 during measurement period **Exclusions:** hospice, palliative care, frailty + advanced illness (≥66)	**MIPS #1 is an inverse measure**. FRDA-related diabetes arises from pancreatic iron deposition and beta-cell dysfunction — code as **E08.–** (diabetes due to underlying condition) rather than E11 to reflect the etiology accurately. Monitor HbA1c at each relevant encounter

MEASURE	STANDARD	APPLICABILITY TO FRDA
Plan All-Cause Readmissions (CMS Star C15; MIPS #479)	**Denominator:** enrollees ≥18 with acute inpatient discharge **Numerator:** unplanned readmission within 30 days (lower is better) **Exclusions:** planned readmissions, hospice, psychiatric	**MIPS #479 is group-level only**; individual PCPs cannot submit it directly. Heart failure and aspiration pneumonia are the primary readmission drivers in FRDA. Omaveloxolone elevates BNP as a class effect without true cardiac decompensation — document drug-induced BNP elevation explicitly in the discharge summary to avoid mislabeled readmissions and unwarranted escalation of heart failure management
Care for Older Adults - Functional Status and Medication Review (HEDIS COA)	**Denominator:** MA enrollees ≥66, continuously enrolled **Numerator:** all 4 sub-measures documented annually: functional status assessment, medication review, pain assessment, advance care planning **Exclusions:** hospice, ESRD	**No single MIPS number maps to COA as a composite**; it remains HEDIS/Stars-only. Record mFARS or SARA score at each visit to fulfill the functional status sub-measure; annual medication reconciliation should note FRDA-specific cautions including CYP3A4 interactions with omaveloxolone

ABBREVIATIONS: FRDA = Friedreich ataxia; HEDIS = Healthcare Effectiveness Data and Information Set; CMS = Centers for Medicare & Medicaid Services; MIPS = Merit-based Incentive Payment System; HbA1c = glycated hemoglobin; BNP = B-type natriuretic peptide; CYP3A4 = cytochrome P450 3A4; mFARS = modified Friedreich Ataxia Rating Scale; SARA = Scale for the Assessment and Rating of Ataxia; COA = Care for Older Adults

QUALITY OUTCOME

Documenting the FRDA subtype and each affected organ system supports appropriate fall screening, glycemic monitoring, and functional assessment. This coding specificity reflects coordinated clinical management rather than administrative documentation alone.

AAVBC PERSPECTIVE

AAVBC encourages primary care providers to treat progressive coordination problems, such as frequent falls, or loss of **deep tendon reflexes,** in a teenager or young adult as an indication

for prompt genetic testing referral, particularly when accompanied by unexplained **hypertrophic cardiomyopathy** or **scoliosis**. The correct diagnostic test is **GAA repeat expansion testing using Triplet Repeat PCR (TP-PCR)**; standard NGS panels frequently miss the causative repeat expansion and should not be substituted. Cardiac workup including **EKG and echocardiogram** should be initiated in parallel with genetic evaluation and should not await neurologist confirmation, as hypertrophic cardiomyopathy is present in up to 80% of patients and is the leading cause of mortality.

AAVBC supports recognition that atypical presentations, including **late-onset Friedreich Ataxia** after age 25 or 40, and a variant in which deep tendon reflexes are retained account for approximately 25% of cases and will be missed by providers relying on the classic pediatric-onset picture.

Beyond diagnosis, AAVBC encourages primary care providers to maintain active longitudinal ownership of the multisystem comorbidity burden, including **cardiomyopathy surveillance**, **diabetes monitoring**, and **orthopedic coordination**, in partnership with neurology and a designated **Ataxia Center of Excellence**. This helps ensure the right patients are identified, appropriately tested, and supported within the right clinical framework.

5 CODING REMINDERS AND CASE EXAMPLES

Coding Specificity

SCENARIO	PREFERRED CODE(S)	SPECIFICITY GUIDANCE
Genetically confirmed Friedreich ataxia	**G11.11** (Friedreich ataxia)	Most specific billable code; includes LOFA, VLOFA, and FARR. Never recode late-onset FRDA as **G11.2**
Testing pending or inconclusive	**G11.10** (Congenital nonprogressive ataxia) or G11.9 (Hereditary ataxia, unspecified)	Temporary only; replace with **G11.11** once the FXN GAA expansion is confirmed
FRDA cardiomyopathy	**I42.2** — Other hypertrophic cardiomyopathy → **I42.0** — Dilated cardiomyopathy; add **I50.x** — Heart failure (unspecified subtype requires further digit specification, e.g., **I50.9** unspecified, **I50.2-** systolic, **I50.3-** diastolic)	Document phenotype and LVEF; never default to **I42.9**; add the heart-failure code alongside, not instead of, the cardiomyopathy code

SCENARIO	PREFERRED CODE(S)	SPECIFICITY GUIDANCE
FRDA-related diabetes	**E08.-** (Diabetes mellitus due to underlying condition) with **G11.11** sequenced first — diabetes secondary to Friedreich ataxia	Diabetes due to an underlying condition, not **E11.-** or **E10.-**; specify complications and insulin use
Neuromuscular scoliosis	**M41.4-** — Neuromuscular scoliosis (site-specific — requires additional digit for spinal region, e.g., **M41.45** thoracolumbar region)	Neuromuscular, not idiopathic **M41.2-**; add the FRDA code as the underlying cause
Dysphagia with aspiration risk	**R13.12** (Dysphagia, oropharyngeal phase)/**R13.13** (Dysphagia, pharyngeal phase) (+ **J69.0** (Pneumonitis due to inhalation of food and vomit (aspiration pneumonia))	Phase-specific rather than **R13.10**; add aspiration pneumonitis when documented

ABBREVIATIONS: FRDA = Friedreich ataxia; FXN = frataxin gene; LOFA = Late-Onset FRDA; VLOFA = Very-Late-Onset FRDA; FARR = FRDA with Retained Reflexes; HF = heart failure; LVEF = left ventricular ejection fraction; HCC = Hierarchical Condition Category

Annual Clinical Review and Confirmation

- Hereditary ataxia is a chronic, lifelong diagnosis, but it must be actively reaffirmed **each year** for the documented complexity to remain accurate
- At the annual visit, restate the confirmed subtype and genetic basis (e.g., "**Friedreich ataxia** confirmed by **FXN GAA repeat expansion**") rather than carrying forward an unspecified code
- Reconcile every active systemic manifestation in the same encounter — **cardiac, endocrine, swallowing, musculoskeletal, urinary, and psychiatric** — since each is coded separately and together they represent the true burden a single neurological HCC cannot convey
- Confirm the status of each problem (**active, stable, resolved**) and the supporting MEAT elements, so the assessment reflects current findings rather than a copied prior note
- Update cardiac and diabetes codes as phenotypes evolve — for example, from **hypertrophic** to **dilated cardiomyopathy**
- All hereditary ataxias map to **HCC 200** (CNA RAF **0.279**), so accurate documentation represents clinical reality and supports coordinated care

Good Documentation - EHR Tips

EHR TIP	HOW IT HELPS	EXAMPLE
FRDA problem-list template	Prompts review of each active system at every visit	Friedreich ataxia **(G11.11)** with linked cardiomyopathy, diabetes, dysphagia, scoliosis entries
Genetic-confirmation SmartPhrase	Keeps the specific etiology in the note to support **G11.11**	'.FRDAconfirm' inserts 'Friedreich ataxia confirmed by FXN GAA repeat expansion'
Omaveloxolone monitoring reminder	Tracks LFTs, BNP, and lipids on schedule	Best-practice alert at baseline and monthly x3 for liver enzymes
Diabetes etiology prompt	Steers coding to **E08.-** rather than **E11.-**	Order-entry note: 'diabetes due to Friedreich ataxia - insulin-first'
Falls and dysphagia screen flags	Documents quality measures and prevents complications	Annual falls-risk and swallowing-screen checkboxes

ABBREVIATIONS: EHR = electronic health record; FRDA = Friedreich ataxia; FXN = frataxin gene; LFT = liver function test; BNP = B-type natriuretic peptide

Brief Case Examples

SUCCESS - Subtype Confirmed, Multisystem Burden Documented

Patient: Female patient, 68, with Very-Late-Onset Friedreich ataxia confirmed by FXN GAA repeat testing[1] after a negative NGS panel.

Documentation: Note records 'Friedreich ataxia (FXN-confirmed) with nonobstructive hypertrophic cardiomyopathy (LVEF 55%, NYHA II) and insulin-treated diabetes due to FRDA;' annual ECG, echocardiogram, HbA1c, swallowing screen, and PHQ-9 documented; omaveloxolone with LFT monitoring noted.

Outcome: Coordinated neurology, cardiology, and endocrinology care; a BNP rise was correctly attributed to omaveloxolone, avoiding an unnecessary admission.

Coding: G11.11 + I42.2 + E08.- documented the full multisystem burden (HCC 200) accurately.

PITFALL - Unspecified Coding and Omitted Complexity

Documentation as written: 'Ataxia, stable. Continue meds.' Coded **G11.9.**

Consequence: Cardiomyopathy, diabetes, and dysphagia went undocumented; the patient was later admitted with aspiration pneumonia and newly recognized heart failure.

RAF/complexity impact: An unspecified code with no comorbidities omitted the multisystem complexity that defines FRDA, so the record understated the patient's true clinical burden.

Fix: Confirm the subtype **(G11.11)**, reconcile and code each active system, and document MEAT for each problem.

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