AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Friedreich Ataxias

Quick Reference Guide

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Table of Contents

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]

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