FACT:

The mFARS Neurological Exam Provides a Detailed Evaluation of a Patient's Status[1]

Friedreich's ataxia (FA) is the most common inherited ataxia.[2] While progression varies by patient, several studies estimate that between 55% and 78% of patients with FA will require a wheelchair within 10 to 15 years after disease onset. For those with more severe FA, this can occur in as few as 3 years.[3]

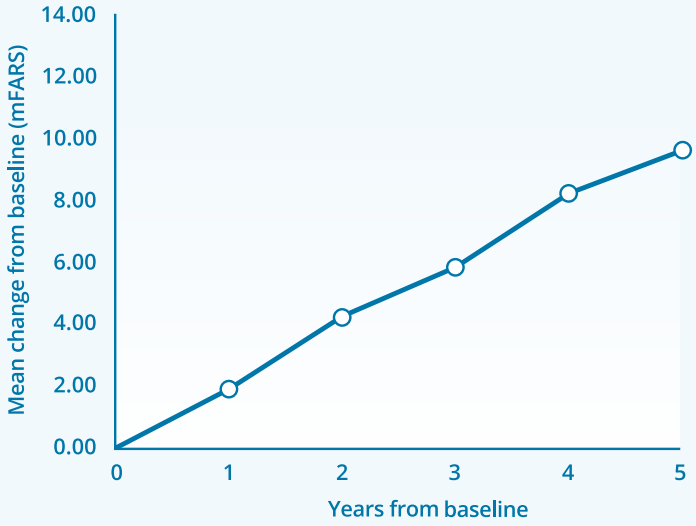
The modified Friedreich's Ataxia Rating Scale (mFARS) is a clinically validated set of assessments that measures FA progression and its impact on a patient's ability to perform activities of daily living.[1] Changes in key mFARS scores help predict time to loss of ambulation and inform management strategy.[4,5]

The mFARS Assesses Current Status and Helps Predict Future Decline[1,4]

4: The number of mFARS evaluation items. Each item is scored to determine the current severity of disability in a specific area[1]

93: The maximum mFARS composite score across all 4 items[1]

65: Average mFARS score where loss of ambulation occurs[6]



Patient mFARS Composite Scores Will Typically Increase ~2 Points per Year

Adapted from: Patel M, et al. *Ann Clin Transl Neurol.* 2016;3(9):684-694. In a multicenter natural history study, 812 diagnosed patients were evaluated annually using multiple tests, including FARS and mFARS. Mean length of GAA triple repeat was 636 and mean age of onset was 13.7 years. Results shown are average annual change from baseline in mFARS composite score for overall cohort.

The mFARS Is Typically Accepted as a Clinical Trial Endpoint

Because of their correlation with disease progression, changes in mFARS scores over time are typically accepted as an endpoint in clinical trials for potential new FA treatments.[1]

A patient's disease severity and rate of progression directly relate to the extent of the genetic triplet-repeat expansion that causes FA. A larger expansion is associated with more serious symptoms and a more rapid decline in function.[3,7]

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The 4 Components of mFARS

The mFARS includes traditional elements of a neuromuscular assessment that specifically focus on a patient's disabilities.

Assessment	Related Daily Activities	Assessment Goals	What This Means for Patients[1]
Bulbar Function	Swallowing or speaking	Measure strength and volume of coughing and clarity of speech.[1,3,7]	Asking the patient to cough 3 times can reveal difficulties with swallowing, catching their breath, or clearing their airways. **Clinical/functional impact:** Risk of respiratory infection. This component also asks the patient to say 2 phrases, such as: "The traffic is heavy today." and "The president lives in the White House." **Clinical/functional impact:** Ability to communicate clearly, a key element in maintaining independence.
Upper Limb Coordination	Brushing teeth, typing, pointing, reaching, or turning a doorknob	Measure motor abilities related to tremors, fine motor coordination, and steadiness of hands and arms.[8,9]	The patient performs finger-to-finger, nose-to-finger, finger-chase, and rapid hand movements. **Clinical/functional impact:** Ability to complete many activities of daily living, including getting dressed and eating, as well as written or electronic communication abilities. These tests can also reveal the presence of tremors.
Lower Limb Coordination	Putting on socks and shoes	Measure coordination of legs and feet.[4]	The patient performs a heel-shin slide and heel-shin tap. **Clinical/functional impact:** Likely rate of disease progression, which is also closely correlated with upright stability results.
Upright Stability	Walking, sitting in a car, standing in line, or showering	Measure loss of ambulation and impaired ability to coordinate voluntary movements (ataxia).[4,10]	The rate of progression is most evident in this component set. Six components evaluate the amount of time a patient can stand or walk in a steady position without listing or needing assistive devices. Three sitting components assess the patient's seated posture. **Clinical/functional impact:** Predicts time to loss of function.

Get tools, information, and resources to help your patients at ThinkFA.com.

References: 1. Rummey C, Corben LA, Delatycki MB, et al. Psychometric properties of the Friedreich Ataxia Rating Scale. *Neurol Genet.* 2019;5(6):371. **2.** National Institute of Neurological Disorders and Stroke. Friedreich Ataxia Fact Sheet. Updated November 15, 2021. Accessed March 16, 2022. https://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Friedreichs-Ataxia-Fact-Sheet. **3.** Parkinson MH, Boesch S, Nachbauer W, Mariotti C, Giunti P. Clinical features of Friedreich's ataxia: classical and atypical phenotypes. *J Neurochem.* 2013;126(suppl 1):103-117. **4.** Rummey C, Farmer JM, Lynch DR. Predictors of loss of ambulation in Friedreich's ataxia. *EClinicalMedicine.* 2020;18:100213. **5.** Pandolfo M. Neurologic outcomes in Friedreich ataxia: Study of a single-site cohort. *Neurol Genet.* 2020;6(3):e415. **6.** Patel M, Isaacs CJ, Seyer L, et al. Progression of Friedreich ataxia: quantitative characterization over 5 years. *Ann Clin Transl Neurol.* 2016;3(9):684-694. **7.** Schulz JB, Boesch S, Bürk K, et al. Diagnosis and treatment of Friedreich ataxia: a European perspective. *Nat Rev Neurol.* 2009;5(4):222-234. **8.** Johns Hopkins Medical. Neurological Exam. Accessed March 16, 2020. https://www.hopkinsmedicine.org/health/conditions-and-diseases/neurological-exam. **9.** de Silva RN, Vallortigara J, Greenfield J, Hunt B, Giunti P, Hadjivassiliou M. Diagnosis and management of progressive ataxia in adults. *Pract Neurol.* 2019;19(3):196-207. **10.** Friedman LS, Farmer JM, Perlman S, et al. Measuring the rate of progression in Friedreich ataxia: implications for clinical trial design. *Mov Disord.* 2010;25(4):426-432.

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