

The modified Friedreich's Ataxia Rating Scale - mFARS

The modified Friedreich's Ataxia Rating Scale – mFARS – is an instrument used to measure neurological function. The exam was specifically developed for FA and includes assessment of neurological signs and symptoms that specifically reflect neural substrates affected in the disease.

Based on a neurological examination, functions from 4 domains are assessed with 18 items yielding a maximum score of 93. The modified version of the FARS (total score of 93) is endorsed for use as primary outcome in clinical trials and it has gained some acceptance with the FDA for that purpose.

In addition, a collaborative initiative with scientists at the Critical Path Institute (C-Path) is working to further increase knowledge and eventually acceptance of the endpoint at regulatory authorities in the United States and Europe. The project is intending to use the Fit-for-Purpose program through the Office of Clinical Pharmacology at FDA, based on the Friedreich's Ataxia Integrated Clinical Database ([FA-ICD](#)), which is part of the rare disease accelerator platform ([RDCA-DAP](#)) created by C-Path.

The FARS score is used in the FACOMS natural history study since 2003 and was published initially in 2005. Revisions thereafter focused on stance abilities and items that directly assess functional, patient relevant abilities, with feedback from the FDA.

Of note, the full-item set of the FARS had 25 items (total score of 125), and while the items used the total score were changed several times based on contemporary knowledge, the complete core-item set was always collected during the exams.

Alongside the neurological exam the concept of Measuring Friedreich's Ataxia also provides other instruments:

1. A Functional Disease Staging (FA-FDS), scored 0 to 6 assessing overall mobility (e.g. 5 being non-ambulatory)
2. An Activities of Daily Living scale (FA-ADL, scored 0 to 36)
3. The Timed 25-foot walk (T25FW)
4. The 9-hole peg board test (9HPT)

Key References on the FARS Score

Subramony SH, May W, Lynch D, Gomez C, Fischbeck K et al. Measuring Friedreich ataxia: Interrater reliability of a neurologic rating scale. *Neurology* 2005;64:1261-1262.

Lynch DR, Farmer JM, Tsou AY, Perlman S, Subramony SH et al. Measuring Friedreich ataxia: complementary features of examination and performance measures. *Neurology* 2006; 66:1711-1716.

[Rummev C, Corben LA, Delatycki MB, Subramony SH, Bushara K et al. Psychometric properties of the Friedreich Ataxia Rating Scale. *Neurology Genetics* 2019; 5:e371](#)

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|-------------------------------------|-----|------|------------------------------------|
| FARS A Bulbar (5) | A3 | (2) | cough |
| | A4 | (3) | speech |
| FARS B Upper Limbs (36) | B1 | (6) | finger-finger |
| | B2 | (8) | nose-finger |
| | B3 | (8) | dysmetria |
| | B4 | (6) | rapid movements |
| | B5 | (8) | finger taps |
| FARS C Lower Limbs (16) | C1 | (8) | heel shin slide |
| | C2 | (8) | heel shin tap |
| FARS E Upright Stability (36) | E1 | (4) | sitting position |
| | E2A | (4) | stance feet apart |
| | E2B | (4) | stance feet apart (eyes closed) |
| | E3A | (4) | stance feet tog. |
| | E3B | (4) | stance feet tog. (eyes closed) |
| | E4 | (4) | tandem stance |
| | E5 | (4) | stance dom. foot |
| | E6 | (3) | tandem walk |
| E7 | (5) | gait | |