

# Friedreich's ataxia CDE Revision History Document

## Contents

|                                |   |
|--------------------------------|---|
| August 2023 Revisions .....    | 3 |
| June 2023 Revisions .....      | 3 |
| August 2022 Revisions .....    | 3 |
| January 2020 Revisions .....   | 4 |
| April 2015 Revisions .....     | 4 |
| March 2015 Revisions .....     | 4 |
| February 2015 Revisions .....  | 4 |
| January 2015 Revisions .....   | 4 |
| November 2014 Revisions .....  | 4 |
| September 2014 Revisions.....  | 4 |
| August 2014 Revisions .....    | 4 |
| July 2014 Revisions .....      | 4 |
| June 2014 Revisions .....      | 4 |
| May 2014 Revisions .....       | 4 |
| April 2014 Revisions.....      | 5 |
| March 2014 Revisions .....     | 5 |
| February 2014 Revisions .....  | 5 |
| January 2014 Revisions .....   | 5 |
| December 2013 Revisions.....   | 5 |
| November 2013 Revisions .....  | 5 |
| July 2013 Revisions .....      | 5 |
| Overview .....                 | 5 |
| Form Status Changes .....      | 5 |
| Detailed Form Revisions: ..... | 7 |

## **Friedreich's ataxia CDE Revision History Document**

|                                |    |
|--------------------------------|----|
| August 2012 Revisions .....    | 10 |
| Overview .....                 | 10 |
| Form Status Changes .....      | 11 |
| Detailed Form Revisions: ..... | 12 |

# Friedreich's ataxia CDE Revision History Document

## August 2023 Revisions

- In the Outcomes and Endpoints; Ataxia and Performance Measures domain; subdomain:
  - The International Cooperative Ataxia Rating Scale (ICARS) was reclassified from Core to Supplemental
  - The Sloan Low Contrast Letter Acuity was reclassified from Core to Supplemental-Highly Recommended
  - 9-Hole Pegboard was renamed as Nine Hole Peg Test
- The FA Highlight Summary and Startup Resources documents were updated to reflect these revisions.

## June 2023 Revisions

- Removed the Friedreich's Ataxia Rating Scale (FARS) Notice of Copyright
- Added the Modified Friedreich's Ataxia Rating Scale (mFARS) Notice of Copyright; Classification: Core to the Outcomes and Endpoints; Ataxia and Performance Measures domain; subdomain.

## August 2022 Revisions

- Added the Listening in Spatialized Noise – Sentences (LiSN-S) Test Guidance document to the Outcomes and End Points/Ataxia Performance and Measures Domain/Subdomain.
- F0923 Prenatal and Perinatal History CDE Details
  - Removed C00311 Birth weight report value
  - Added C59019 Birth weight measurement
- F0940 Activities of Daily Living and Gait CRF and CDE Details
  - Added Permissible Value (PV) 'Without any difficulty' to CDEs C10692 Speech level status; C10693 Swallowing level status; C10694 Cutting food or use of items functioning level status; C10696 Personal hygiene level status; C10698 Walking ability status; C10699 Sitting ability status; C10700 Bladder control status and C10695 Dressing level status to PV Set IDs.
  - Revised 'Normal' to 'Without any difficulty' in the PV Subsets for these CDEs and updated 'Normal' to 'Without any difficulty' for Questions 1-5, 7-9 on the F0940 Activities of Daily Living and Gait CRF.
  - Added Permissible Value (PV) 'Never' to CDE C10697 Fall rate.
  - Revised 'Normal' to 'Never' in the PV Subset for this CDE C10697 and for Question 6 on the F0940 Activities of Daily Living and Gait CRF.

# **Friedreich's ataxia CDE Revision History Document**

## **January 2020 Revisions**

- General Core CRF updated to replace 'Gender' question with 'Sex assigned at birth' and 'Gender identity'.
- Two new CDEs added to CRF: C58676 (Birth sex assigned type) and C58677 (Gender identity type). C00035 has been removed.
- Start-Up document updated to reflect these changes.

## **April 2015 Revisions**

No changes.

## **March 2015 Revisions**

- The Start-Up Resource Listings were revised.

## **February 2015 Revisions**

No changes.

## **January 2015 Revisions**

- Changes to FA Acknowledgements page regarding Dr. Petra Kaufmann.

## **November 2014 Revisions**

No changes.

## **September 2014 Revisions**

No changes.

## **August 2014 Revisions**

No changes.

## **July 2014 Revisions**

No changes.

## **June 2014 Revisions**

No changes.

## **May 2014 Revisions**

No changes.

# Friedreich's ataxia CDE Revision History Document

## April 2014 Revisions

No changes.

## March 2014 Revisions

No changes.

## February 2014 Revisions

No changes.

## January 2014 Revisions

- New Notices of Copyright loaded for FA.

## December 2013 Revisions

- Updated highlight summaries and start-up resource documents.

## November 2013 Revisions

No changes.

## July 2013 Revisions

### Overview

There have been changes to the content and structure of the Friedreich's Ataxia (FA) CDE materials in Version 1.2 compared to Version 1.1, including the following:

### Form Status Changes

#### New Forms:

Acoustic Analysis of Speech

Barthel Index

Functional Independence Measure (FIM)

Jebsen-Taylor Hand Function Test

PaTaKa Speech Test

Assessment of Intelligibility of Dysarthric Speech (AIDS)

Boston Diagnostic Aphasia Examination – Third Edition (BDAE-3)

## **Friedreich's ataxia CDE Revision History Document**

Delis-Kaplan Executive Function System (D-KEFS)

International Cooperative Ataxia Rating Scale (ICARS)

Phonemic Verbal Fluency

Pediatric Quality of Life Inventory (Peds QL)

SF-10 Health Survey for Children

### **Updated Forms:**

Notice of Copyright Created (NOC) (Will replace previously existing forms)

Bladder Control Scale (BLCS) (Component of Multiple Sclerosis Quality of Life)

Bowel Control Scale (BWCS) (Component of Multiple Sclerosis Quality of Life)

Friedreich's Ataxia Impact Scale (FAIS)

Friedreich's Ataxia Rating Scale

Impact of Visual Impairment Scale (IVIS)

Modified Fatigue Impact Scale (MFIS)

MOS Pain Effects Scale (PES) (Component of Multiple Sclerosis Quality of Life)

Nine-Hole Peg Test

Scale for the Assessment and Rating of Ataxia (SARA)

Sloan Low Contrast Letter Acuity

Tardieu Scale

Timed 25-Foot Walk (T25FW)

36-Item Short Form Health Survey (SF-36)

### **Removed Forms:**

## Friedreich's ataxia CDE Revision History Document

None

### Detailed Form Revisions:

Additional specific changes to the CRF modules are detailed in the table included on the subsequent page.

#### 1 Friedreich's Ataxia Revision History Table

| CRF Module/<br>Guideline   | Current<br>Version | Previous<br>Version | Change<br>Classification | Change Type            | Description of<br>Change  |
|--|--------------------|---------------------|--------------------------|------------------------|---|
| Acoustic<br>Analysis of<br>Speech                                  | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Barthel Index  | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Functional<br>Independence<br>Measure                              | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Jebsen-Taylor<br>Hand Function<br>Test                             | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| PaTaKa Speech<br>Test  | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Assessment of<br>Intelligibility of<br>Dysarthric<br>Speech (AIDS) | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |

## Friedreich's ataxia CDE Revision History Document

| CRF Module/<br>Guideline   | Current<br>Version | Previous<br>Version | Change<br>Classification | Change Type            | Description of<br>Change  |
|--|--------------------|---------------------|--------------------------|------------------------|---|
| Boston<br>Diagnostic<br>Aphasia<br>Examination –<br>Third Edition<br>(BDAE-3)                    | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Delis-Kaplan<br>Executive<br>Function<br>System (D-<br>KEFS)                                     | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| International<br>Cooperative<br>Ataxia Rating<br>Scale (ICARS)                                   | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Phonemic<br>Verbal Fluency   | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Pediatric<br>Quality of Life<br>Inventory<br>(Peds QL)   | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| SF-10 Health<br>Survey for<br>Children   | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |
| Bladder<br>Control Scale<br>(BLCS)<br>(Component of<br>Multiple<br>Sclerosis<br>Quality of Life) | 2.0                | 1.2                 | Major                    | Notice of<br>Copyright | A Notice of<br>Copyright was<br>created for this<br>form and will<br>replace the<br>existing form |



## Friedreich's ataxia CDE Revision History Document

| CRF Module/<br>Guideline  | Current<br>Version | Previous<br>Version | Change<br>Classification | Change Type         | Description of<br>Change   |
|---|--------------------|---------------------|--------------------------|---------------------|--|
| Bowel Control Scale (BWCS)<br>(Component of Multiple Sclerosis Quality of Life)   | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Friedreich's Ataxia Impact Scale (FAIS)   | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Friedreich's Ataxia Rating Scale  | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Impact of Visual Impairment Scale (IVIS)  | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Modified Fatigue Impact Scale (MFIS)  | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| MOS Pain Effects Scale (PES)<br>(Component of Multiple Sclerosis Quality of Life) | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Nine-Hole Peg Test  | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |

## Friedreich's ataxia CDE Revision History Document

| CRF Module/<br>Guideline                             | Current<br>Version | Previous<br>Version | Change<br>Classification | Change Type         | Description of<br>Change   |
|--|--------------------|---------------------|--------------------------|---------------------|--|
| Scale for the Assessment and Rating of Ataxia (SARA) | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Sloan Low Contrast Letter Acuity                     | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Tardieu Scale  | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| Timed 25-Foot Walk (T25FW)                           | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |
| 36-Item Short Form Health Survey (SF-36)             | 2.0                | 1.2                 | Major                    | Notice of Copyright | A Notice of Copyright was created for this form and will replace the existing form |

### August 2012 Revisions

#### Overview

There have been changes to the content and structure of the Friedreich's Ataxia (FA) CDE materials in Version 1.1 compared to Version 1.0, including the following:

- Every attempt was made to remove duplication and redundancy in the FA CDE(s)
- The FA CDE(s) were harmonized with other large national and international standardization initiatives and efforts [e.g., International Conference on Harmonization (ICH), Logical Observation Identifiers Names and Codes (LOINC), International Classification of Diseases (ICD), Unified Code for Units of Measure (UCUM)]

## **Friedreich's ataxia CDE Revision History Document**

- The majority of element names and definitions were improved.
- The permissible values and references of some elements were revised; and
- The version numbers of all forms (excluding standardized scales/instruments) were updated.
- Several guideline documents were reorganized and consolidated to make it easier to understand which standardized instruments/scales are recommended, though the recommendations themselves remained unchanged.
- Some of the forms and guideline documents were re-categorized under a different domain or subdomain to be consistent with other NINDS CDEs.
- The corresponding data dictionaries (i.e., CDE Detailed Report) have been modified in accordance with the form changes.

### **Form Status Changes**

#### **New Forms:**

- Medical History of Friedreich's Ataxia (formerly part of the "Adult Medical History and Prior Health Status" CRF)
- Medical History (formerly part of the "Adult Medical History and Prior Health Status" CRF)
- Pediatric Developmental History (formerly part of the "Pediatric Medical History" CRF)
- Physical Examination
- Pregnancy History (formerly part of the "Behavioral and Social History" CRF)
- Prenatal and Perinatal History (formerly part of the "Pediatric Medical History" CRF)
- Prior and Concomitant Medications (created using the elements from the "Medication History" CRF)
- Social Status (formerly part of the "Demographics and Social Status CRF")

#### **Updated Forms:**

- Behavioral History
- Demographics
- Family History
- History Data Source and Reliability

## Friedreich's ataxia CDE Revision History Document

- Laboratory Tests
- Plasma Collection Guidelines
- Urine Collection Guidelines

### Removed Forms:

- Adult Medical History and Prior Health Status
- Medication History
- Pediatric Medical History

### **Detailed Form Revisions:**

Additional specific changes to the CRF modules are detailed in the table included on the subsequent page

### **2 Friedreich's Ataxia Revision History Table**

| CRF Module/<br>Guideline | Current<br>Version | Previous<br>Version | Change<br>Classification | Change<br>Type    | Description of Change  |
|--------------------------|--------------------|---------------------|--------------------------|-------------------|--|
| All                      | 1.1                | 1.0                 | Minor                    | Modified<br>CDEs  | Nearly all CDE names and definitions were updated to achieve greater consistency across all NINDS CDE(s) and for clarity                                 |
| Various                  | 1.1                | 1.0                 | Minor                    | Deleted<br>CDEs   | The following types of CDEs were deleted: CDEs redundant with other CDE(s); CDEs specifying unit; and CDE(s) specifying an "other, specify" response     |
| Various                  | 1.1                | 1.0                 | Minor                    | Modified<br>CDEs  | For a number of CDEs, "Unknown" was added to the permissible value set and in some cases "Unknown" replaced "Unknown/Don't Know"                         |
| Various                  | 1.1                | 1.0                 | Minor                    | Modified<br>Forms | For forms with Core items, the Core CDE(s) were identified with an asterisk and the following was added as a footnote: "*Element is classified as Core". |
| Various                  | 1.1                | 1.0                 | Minor                    | Modified<br>Forms | Instructions were updated to include additional details.   |
| Various                  | 1.1                | 1.0                 | Minor                    | Modified<br>Forms | Friedreich's Ataxia was removed from the header of all forms.  |

## Friedreich's ataxia CDE Revision History Document

| CRF Module/<br>Guideline | Current<br>Version | Previous<br>Version | Change<br>Classification | Change<br>Type   | Description of Change   |
|--------------------------|--------------------|---------------------|--------------------------|------------------|---|
| Behavioral<br>History    | 1.1                | 1.0                 | Minor                    | Modified<br>Form | The form title was changed from "Behavioral and Social History" to "Behavioral History".  |
| Behavioral<br>History    | 1.1                | 1.0                 | Minor                    | Modified<br>CDEs | Permissible values were modified for the following CDEs: "Tobacco current use indicator"; "Tobacco prior use indicator"; "Tobacco use started age value"; "Tobacco use stopped age value"; "Tobacco product used type"; "Tobacco cigarettes smoked daily average number"; "Drug or substance current illicit use indicator"; "Drug or substance illicitly used category"  |
| Behavioral<br>History    | 1.1                | 1.0                 | Minor                    | Added<br>CDEs    | The following CDE was added: "Behavioral history assessment date and time"  |
| Behavioral<br>History    | 1.1                | 1.0                 | Minor                    | Modified<br>Form | Instructions were added for the following CDEs: "Behavioral history assessment date and time"; "Tobacco current use indicator"; "Tobacco prior use indicator"; "Tobacco use started age value"; "Tobacco use stopped age value"; "Tobacco product used type"; "Tobacco cigarettes smoked daily average number"; "Alcohol current use indicator"; "Alcohol prior use indicator"; "Alcohol use started age value"; "Alcohol use stopped age value"; "Alcohol use frequency"; "Alcohol drinking day average drinks consumed range"; "Alcohol consume six or more drinks frequency"; "Alcohol use related to hospitalization indicator"; "Drug or substance current illicit use indicator"; "Drug or substance illicitly used category" |

## Friedreich's ataxia CDE Revision History Document

| CRF Module/<br>Guideline                     | Current<br>Version | Previous<br>Version | Change<br>Classification | Change<br>Type     | Description of Change  |
|--|--------------------|---------------------|--------------------------|--------------------|--|
| Demographics                                 | 1.1                | 1.0                 | Minor                    | Modified<br>CDEs   | The following definition and/ or permissible values were updated for the following CDEs: "Gender type"; "Ethnicity USA category"; "Race USA category"  |
| Demographics                                 | 1.1                | 1.0                 | Minor                    | Deleted<br>CDEs    | The following CDE was deleted: "Hand preference type" (moved to Physical Examination CRF). The CDE(s) on this form under the heading of "Social Status Information" were removed and moved to the Social Status CRF. |
| Demographics                                 | 1.1                | 1.0                 | Minor                    | Modified<br>Form   | Instructions were updated.   |
| Family History                               | 1.1                | 1.0                 | Minor                    | Added<br>CDEs      | The following CDE was added: "Family history medical condition relative type"  |
| History Data<br>Source and<br>Reliability    | 1.1                | 1.0                 | Minor                    | Modified<br>Form   | The name of the form was changed from "Data Source and Reliability" to "History Data Source and Reliability".  |
| Laboratory<br>Tests                          | 1.1                | 1.0                 | Minor                    | Added<br>CDE(s)    | The following CDE was added: "Lab test performed indicator"  |
| Laboratory<br>Tests                          | 1.1                | 1.0                 | Minor                    | Modified<br>CDE(s) | Permissible values were modified for the following CDEs: "Lab test result status"; "Lab test abnormality significance type"  |
| Laboratory<br>Tests                          | 1.1                | 1.0                 | Minor                    | Modified<br>Form   | Instructions were updated.   |
| Medical History<br>of Friedreich's<br>Ataxia | 1.0                | N/A                 | Minor                    | New<br>Form        | CDEs from the Adult Medical History and Prior Health Status CRF under the heading of "Medical History" were moved to this new form.  |
| Medical History                              | 1.0                | N/A                 | Minor                    | New<br>Form        | CDEs from the Adult Medical History and Prior Health Status CRF related to the participant's/subject's overall medical history and not his/her medical history specific to Friedreich's Ataxia were moved to         |

## Friedreich's ataxia CDE Revision History Document

| CRF Module/<br>Guideline                | Current<br>Version | Previous<br>Version | Change<br>Classification | Change<br>Type   | Description of Change  |
|---|--------------------|---------------------|--------------------------|------------------|--|
|   |                    |                     |                          |                  | this new form.   |
| Pediatric<br>Developmental<br>History   | 1.0                | N/A                 | Minor                    | New<br>Form      | CDEs from the Pediatric Medical History CRF under the heading of "Pre-Morbid Developmental Status and Academic Function" were moved to this form.  |
| Pregnancy<br>History                    | 1.0                | N/A                 | Minor                    | New<br>Form      | CDEs from the Behavioral and Social History CRF under the heading of "Pregnancy History" were moved to this form.  |
| Physical Exam                           | 1.0                | N/A                 | Major                    | New<br>Form      | The Physical Exam form was added to the Friedreich's Ataxia recommendations. All CDEs on this form are new with the exception of "Hand preference type".   |
| Plasma<br>Collection                    | N/A                | 1.0                 | Major                    | Modified<br>Form | This document is no longer associated with CDEs and serves as a guideline and not a CRF. The name of the document was changed from "Plasma Collection" to "Plasma Collection Guidelines".  |
| Prenatal and<br>Perinatal<br>History    | 1.0                | N/A                 | Minor                    | New<br>Form      | CDEs from the Pediatric Medical History CRF under the headings of "Delivery History" and "Prenatal History" were moved to this form.   |
| Prior and<br>Concomitant<br>Medications | 1.0                | N/A                 | Major                    | New<br>Form      | CDEs from the Medication History CRF were moved to this form. Several of the CDEs on this form are new.  |
| Social Status                           | 1.0                | N/A                 | Minor                    | New<br>Form      | The following CDEs were removed from the Demographics and Social Status CRF and added to this form: "Education level USA type"; "Employment status"; "Full-time employment status"; "Marital or partner status"; "Living with persons number"; "Living with person relationship type"; "Paid assistance per day hours number"; "Unpaid assistance per day hours number"; "In-home assistance for |

## Friedreich's ataxia CDE Revision History Document

| CRF Module/<br>Guideline    | Current<br>Version | Previous<br>Version | Change<br>Classification | Change<br>Type | Description of Change   |
|-----------------------------|--------------------|---------------------|--------------------------|----------------|---|
|                             |                    |                     |                          |                | remembering, decision making, or judgment duration"; "Family income supported persons number"   |
| Urine Collection Guidelines | N/A                | 1.0                 | Major                    | Modified Form  | This document is no longer associated with CDEs and serves as a guideline and not a CRF. The name of the document was changed from "Urine Collection" to "Urine Collection Guidelines". |