

Friedreich's Ataxia Rating Scale (FARS) and Modified FARS (mFARS)

Test Description

The FARS is a neurological rating scale, developed to measure neurological function in FA. It is a composite measure designed to reflect specific neural substrates affected by FA¹⁻³. The FARS is a five-subscale assessment (sections A to E), measuring bulbar function, upper limb coordination, lower limb coordination, peripheral nervous system, and upright stability. The modified FARS (mFARS) has better psychometric properties and shortens the bulbar subscale to 2 items (from 4) and excludes the 5 items in the peripheral nerve subscale⁴.

This document provides *updated* (March 2023) instructions for performing assessments so that the mFARS can be learned and implemented at more research sites. The scoring is unchanged from the original. The mFARS testing administration order suggested has been arranged to minimize the transfers that need to be performed by the participant.

Of note, this does not include the FA-Activities of Daily Living (FA-ADL) or the FA-Functional Disability Score (FA-FDS), as these are separate assessments.

Test Guidelines

The mFARS should only be completed by a trained examiner, preferably a neurologist. Each study site should strive to have one primary examiner and at least one alternate examiner for any FA clinical research study.

Training will occur annually at a meeting of the FA-GCC. Trained examiners will be required to sign-off on a careful reading and understanding of this document in addition to attending the training meeting.

As this is an assessment of function, it is suggested to perform this assessment at a similar time and in the same order related to other study assessments. In practice, participants should be instructed to get a good night's sleep, eat breakfast, arrive hydrated, and wear comfortable clothing that allow examination to be performed. Participants should be instructed not to exercise or walk more than necessary the morning of testing (e.g., wheelchair or hospital transport chair should be offered if there are long distances participant between parking and/or exam locations).

Scoring

Scoring is based on a composite score of all five subscales with a maximum score of 93 points for mFARS and 125 for FARS. Scores are graded according to each item's criteria with increasing number indicating a higher severity/worse function. The total mFARS/FARS scores for each subscale are as follows:

Subscale	mFARS Scoring (points)	FARS Scoring (points)
A. Bulbar	5	11
B. Upper Limb Coordination	36	36
C. Lower Limb Coordination	16	16
E. Upright Stability	36	36
D. Peripheral Nervous System	-	26
Total Scores	93	125

It is recommended that CRF only capture the score of each individual assessment. The subscale and overall scores should be generated by the database to avoid human error.

Equipment

- Stopwatch
- Colored tape for marking the floor
- Stryker Prime Transport Chair or equivalent stable examination chair, including patient's own adaptive system
- Adjustable Plinth or equivalent exam room table or low foot stool
- If performing FARS: subscale D you will need a 128 cps tuning fork and reflex hammer

The following sequence is to be completed for the mFARS administration.

Modified Friedreich's Ataxia Rating Scale (mFARS)

A. Bulbar, Items A 3 – 4*

Position	Participant is in an upright seated position with 90 degrees of hip flexion, 90 degrees of knee flexion, and feet supported.
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*Naming from original FARS exam; mFARS omits A1 or A2

A. BULBAR					
3. Cough					
Instruction	Cough as hard as you can. Three times. Accept the strongest attempt.				
Assessment Detail	Listen to the strength of the cough. <i>*Increments of 0.5 may be used if examiner feels an item falls between two severities.</i>				
Score*	0.0	0.5	1.0	1.5	2.0
Scoring Specifics	Normal	Depressed		Totally or Nearly Absent	

A. BULBAR							
4. Speech							
Instruction	Ask the participant to read or repeat the following two sentences: A. The President lives in the White House. B. The traffic is heavy today.						
Assessment Detail	<i>*Increments of 0.5 may be used if examiner feels an item falls between two severities.</i>						
Score*	0.0	0.5	1.0	1.5	2.0	2.5	3.0
Scoring Specifics	Normal	Mild (all or most words are understandable)		Moderate (most words are not understandable)		Severe (no or almost no useful speech)	

B. Upper Limb Coordination, Items B 1 – 5

Position	Participant is in an upright seated position with 90 degrees of hip flexion, 90 degrees of knee flexion, and feet supported. If a participant cannot sit unsupported in the defined chair, they may stay seated in their adapted seating system.
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B. UPPER LIMB COORDINATION							
1. Finger to Finger Test							
Instruction	Assist participant into a position of shoulder abduction so that elbows reach the horizontal plane. Elbows are at 75-90 degrees of flexion and hands are 25 cm from the sternum. While supporting their arms, have them touch their index fingers together and then move them 2.5 cm apart. Ask them to hold this position and remove your support. Downward drift of elbows is allowed. Observe for 10 seconds.						
Assessment Detail	Score amplitude of finger oscillations. If elbows require contact with body or surface then they score a 3.0. <i>Increments of 0.5 may be used if examiner feels an item falls between two severities. Test is performed and scored for each limb.</i>						
Score – 2 separate scores for right and left	0.0	0.5	1.0	1.5	2.0	2.5	3.0
Scoring Specifics	Normal (0 – 0.5 cm)	Mild oscillations of finger (0.5 – 2.0 cm)	Moderate oscillations of finger (2.0 – 6.0 cm)	Severe oscillations of finger (greater than 6.0 cm) and/or elbows require contact with body or surface, unable to perform task			

B. UPPER LIMB COORDINATION					
2. Nose - Finger Test					
Instruction	Examiner holds index finger at 90% reach of participant and test 3 nose-finger-nose trials; assess participant's finger path through a straight on view. <i>Test is performed and scored for each limb.</i>				
Assessment Detail	Assess kinetic or intention tremor during and towards the end of the movement.				
Score – 2 separate scores for right and left	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	None	Mild (less than 2.0 cm amplitude)	Moderate (2.0 – 6.0 cm amplitude)	Severe (greater than 6.0 cm)	Too poorly coordinated to perform task

B. UPPER LIMB COORDINATION

3. Dysmetria Test

Instruction	Examiner explains to participant that they need to reach out and touch the tip of examiner’s index finger as it moves to four corners of an imaginary square, going back and forth from participant’s chin to examiner’s finger. The imaginary square is about 25cm each side and should be about 50% distance from the full reach of the patient’s arm. This is repeated 8 times as the examiner verbally cues the participant to touch his/her finger as it moves in a clockwise direction, making 2 complete navigations of the square. Test is performed at a fast pace. The examiner assesses the participant’s forward reach and tracks the accuracy of the movement. <i>Test is performed and scored for each limb.</i>				
Assessment Detail	Assess dysmetria, tip of finger is defined as any part of the examiners DIP joint (i.e., inaccuracy of reaching the target – examiner’s finger). There is no penalty for tremor.				
Score - 2 separate scores for right and left	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	None (no misses)	Mild (1 – 2 misses)	Moderate (misses 3-5 times)	Severe (misses 6-8 times)	Too poorly coordinated to perform task

B. UPPER LIMB COORDINATION

4. Rapid Alternating Movements of Hands

Instruction	Stopwatch required. Have participant flex their elbow so their forearm is 15 cm above the thigh and forearm in a pronated position (palm facing up). Demonstrate forearm pronation/supination and explain that the movement is like placing their hand on a doorknob and turning it back and forth. Cue participant that they should not brace their elbow to their side as this is a compensation. Allow participant to practice as you count out loud. Instruct participant to complete 10 cycles as quickly as possible. 1 cycle is a full supination and pronation movement. Use a stopwatch to measure time to complete 10 cycles. The examiner keeps count and says “go” and “stop.” <i>Test is performed and scored for each limb.</i>					
Assessment Detail	Assess rate, rhythm, body position, and accuracy. If time to completion is >7.0 seconds then 0.5 is added to the score.					
Score - 2 separate scores for right and left	0.0	0.5	1.0	1.5	2.0	2.5 3.0
Scoring Specifics	Normal	Mild (slightly irregular or slowed) but wrist and elbow remain in relatively fixed position away from the torso		Moderate (irregular and slowed) or participant compensates by bracing elbow on their trunk or there is noted elbow excursion during maneuver		Too poorly coordinated to perform task

B. UPPER LIMB COORDINATION

5. Finger Taps

Instruction	Stopwatch required. Position participant with shoulder abducted to horizontal plane and elbows flexed at 75 to 90 degrees. Ask the participant to tap on the thumb crease (DIP joint) with the tip of their index finger with a movement amplitude of at least 1.0 cm. Demonstrate 15 reps as fast as possible. Demonstrate an excursion of at least 1 cm. Cue participant that you will tell them when to start and stop and they should perform task as quickly as possible. Use stopwatch to measure time as you count for the participant. The examiner keeps count and says “go” and “stop.” <i>Test is performed and scored for each limb.</i>				
Assessment Detail	Each miss of the thumb crease is distinguished as a mistake. <i>If time > 6 seconds, add 1 to score.</i>				
Score - 2 separate scores for left and right	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	None	Mild (missed 1-3 times)	Moderate (missed 4-9 times)	Severe (missed 10-15 times)	Cannot perform the task and/or unable to maintain start position
Notes	If they have long fingernails they should be cued to use the tip of their 1 st finger to touch the thumb crease and not use the nail.				

C. Lower Limb Coordination, Items C 1 & 2

Position	Participant is in a supine position on the plinth with their head elevated for visualization of the lower extremities. The participant should lie with knees extended and kneecaps/toes pointing upward. If the participant has difficulty transferring to an exam table/chair this can be done in their adapted seating system with a small stool place in front for their legs to be extended out in front of them with feet supported.
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C. LOWER LIMB COORDINATION

1. Heel Along Shin Slide

Instruction	Demonstrate this maneuver by lifting the participant’s lower leg and guiding it through movement of the heel down the shin. Cue them to watch their heel, touch heel to the opposite leg (just under kneecap), slide heel on the tibia to the ankle, take heel off ankle, place heel down onto plinth or stool, and bring heel back to the contralateral shin- just below patella. Ask them to repeat 3 cycles of this movement at moderate speed- one second per excursion of hip flexion or extension. <i>Test is performed and scored for each limb.</i>				
Assessment Detail	Assessing contact with shin and smoothness of the movement.				
Score - 2 separate scores for right and left	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	Normal (stays on shin)	Mild (abnormally slow, tremulous but contact is maintained)	Moderate (goes off shin less than or equal to 3 times during 3 cycles)	Severe (goes off shin 4 or more times during 3 cycles)	Too poorly coordinated to perform task or cannot assume start position (due to contractures or other impairments).

C. LOWER LIMB COORDINATION

2. Heel to Shin Tap

Instruction	The participant identifies a comfortable spot on their mid-shin as target. Under visual control, participant taps heel on the target on the opposite shin 8 times on each side from about 15-25 cm above from the shin, one at a time. Examiner keeps count and says “go” and “stop.” <i>Test is performed and scored for each limb.</i>				
Assessment Detail	The number of times the heel misses the selected spot is assessed.				
Score - 2 separate scores for each limb	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	Normal (stays on target)	Mild (misses shin 1 to 2 times)	Moderate (misses shin 3-5 times)	Severe (goes off shin more than 5 times)	Too poorly coordinated to perform task or cannot assume start position due to contractures or other impairments.

E. Upright Stability, Items E 1 – 7

- Participant should be **barefoot**; however, if they refuse, they can still do task but record if barefoot or footwear. Record dominant foot, type of flooring, if braces are worn and always perform with same/similar future assessments.
- Test to be performed on the same hard, stable surface for all visits. Preferably, a non-carpeted surface or on a clear hard plastic office chair mat over the carpeted surface for items 1-5.
- The participant should be given the encouragement and support to attempt all the tasks. Don’t assume if the participant arrives to the exam using a mobility device or wheelchair that they are unable to stand or walk.
- Upright stance tests should be performed in front of the adjustable mat table or in front of a secure chair or wheelchair to provide something for the participant to grab if they become off balance or feel they might fall. Study personnel should be close to the subject if needed. If the evaluator feels there is a safety risk, then a test should not be performed and a maximum score applied for all trials of the test.
- Stance assessments will be done with feet apart approximately 20cm. A horizontal tape can be applied to the floor for consistency of measurement and to provide participant with orientation for foot placement. Toes pointed forward, and feet as flat on the floor as possible. Stance with feet together assessments necessitate that the medial border of the feet be touching at the MTP joints and the heels.
- If the participant cannot perform the first stance test (feet apart eyes open) without assistance (holding examiner or table or walker) then a score of 4 is given for all 3 trials of this test and none of the other stance tests are attempted with max score of 4 applied for all trials of all tests (2b, 3a, 3b, and 5). Tandem stance should still be attempted.
- If the participant cannot assume the relevant position for 3a (feet together eyes open) without assistance (holding examiner or table or walker) then a score of 4 is given for all 3 trials of this test and 3b). Tandem stance should still be attempted.
- For all the stance items (2a, 2b, 3a, 3b, 4, & 5) the score for each attempt is captured on the CRF even if the attempt is not performed. The score for the item is the average of the 3 scores (it is recommended that this average is generated by the database when score for the 3 attempts are entered).
- Mark off 20 cm with tape on the floor for stance tasks and 8 meter or 25 foot distance in a hallway or long room for gait items. Record which distance was used. Study personnel should ensure that area has been cleared of clutter or obstacles to reduce fall risk and should be close to the subject. If the evaluator feels there is a safety risk then a test should not be performed and a maximum score applied.

E. UPRIGHT STABILITY

1. Sitting Posture

Position	Participant is in an upright seated position on the edge of the plinth or chair with back unsupported. If participant is not able to transfer from personal adapted seating device, see if they can scoot forward a few inches so that back is unsupported. There should be 1-2 inches of space between the edge of seating platform and the back of their lower leg. The participant should sit with 90 degrees of hip flexion and 90 degrees of knee flexion. The participant will fold their arms across their chest so hands rest on their elbows. If a participant cannot sit unsupported in the defined position, they receive a score of a 4.				
Instruction	Place examiner in front of participant and have an assistant stand behind or to one side of the participant to assist if participant begins to lose their balance.				
Assessment Detail	Observe for 30 seconds.				
Score	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	Normal	Mild oscillations of head/trunk	Moderate oscillations of head/trunk; requires supervision for safety, but no contact guard assistance	Severe oscillations of head/trunk; needs contact to minimal assistance of 1-2 people.	Requires moderate to maximal support of one to two people, or cannot assume start position.

E. UPRIGHT STABILITY

2a. Stance – Feet Apart – Eyes Open

Position	Participant is to stand with medial border of their feet 20cm apart with toes pointing forward along a horizontal tape line on the floor. The MTP joints should be on the horizontal line. The participant will have their arms and hands by their sides or in a position of their choosing during testing with visual gaze fixed at a spot of their choosing.				
Instruction	Stopwatch required. Instruct the participant that you will be timing their ability to stand in this position. Have an examiner and an assistant on each side of the participant to catch them if they lose their balance. Use a stopwatch to measure up to 3 attempts.				
Assessment Detail	As soon as the participant is able to exceed 60 seconds (on the first or second trial), no subsequent trials are required and score of zero is recorded for the remaining trial(s).				
Score	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	1 minute or longer	Less than 1 minute, greater than 45 seconds	Less than 45 seconds, greater than 30 seconds	Less than 30 seconds, greater than 15 seconds	Less than 15 seconds or needs hands held by assistant/device, or cannot assume start position.
Notes	Participant can seek support in between trials; holding on to the table, chair or examiner.				

E. UPRIGHT STABILITY					
2b. Stance – Feet Apart – Eyes Closed					
Position	Same as for item 2a.				
Instruction	Stopwatch required. Inform the participant that they are going to do the same standing task but with their eyes closed. When participant feels ready, say “go” to have them close eyes. Have the examiner and an assistant stand on each side of participant to watch their eyes and assist if participant begins to sway. Stop timing if the participant blinks or opens eyes. Use a stopwatch to measure up to 3 attempts.				
Assessment Detail	As soon as the participant is able to exceed 60 seconds (on the first or second trial), no subsequent trials are required and score of zero is recorded for the remaining trial(s).				
Score	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	1 minute or longer	Less than 1 minute, greater than 45 seconds	Less than 45 seconds, greater than 30 seconds	Less than 30 seconds, greater than 15 seconds	Less than 15 seconds or needs hands held by assistant/device
Notes	Participant can seek support in between trials; holding on to the table, chair or examiner.				

E. UPRIGHT STABILITY					
3a. Stance – Feet Together – Eyes Open					
Position	Participant is to stand with medial border of feet touching at both the MTP joints and heels. If the participant is “knock-kneed” or has other orthopedic issue that prevents feet from touching at front and back then having feet touching at one location is sufficient. The participant will have their arms and hands by their sides or in a position of their choosing during testing with visual gaze fixed at a spot of their choosing.				
Instruction	Stopwatch required. Instruct the participant that you will be timing their ability to stand in this position. Have the examiner and an assistant stand on each side of participant to assist if participant begins to sway. Use a stopwatch to measure up to 3 attempts.				
Assessment Detail	As soon as the participant is able to exceed 60 seconds (on the first or second trial), no subsequent trials are required and score of zero is recorded for the remaining trial(s).				
Score	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	1 minute or longer	Less than 1 minute, greater than 45 seconds	Less than 45 seconds, greater than 30 seconds	Less than 30 seconds, greater than 15 seconds	Less than 15 seconds or needs hands held by assistant/device, or cannot assume start position
Notes	Participant can seek support in between trials; holding on to the table, chair or examiner.				

E. UPRIGHT STABILITY					
3b. Stance – Feet Together – Eyes Closed					
Position	Same as for 3a.				
Instruction	Stopwatch required. Inform the participant that they are going to do the same standing task but with their eyes closed. When participant feels ready, say “go” to have them close eyes. Have examiner and an assistant stand on each side of participant to watch their eyes and assist if participant begins to sway. Stop timing if the participant blinks or opens eyes. Use a stopwatch to measure up to 3 attempts.				
Assessment Detail	As soon as the participant is able to exceed 60 seconds (on the first or second trial), no subsequent trials are required and score of zero is recorded for the remaining trial(s).				
Score	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	1 minute or longer	Less than 1 minute, greater than 45 seconds	Less than 45 seconds, greater than 30 seconds	Less than 30 seconds, greater than 15 seconds	Less than 15 seconds or needs hands held by assistant/device, or cannot assume start position.
Notes	Participant can seek support in between trials; holding on to the table, chair or examiner.				

E. UPRIGHT STABILITY					
4. Tandem Stance					
Position	Assist participant into a tandem stance position with their dominant foot in back and non-dominant foot in front. The dominant foot is the one they feel they balance the best upon when asked to stand on one leg. The toe of the dominant foot should touch the heel of the non-dominant foot. Document which leg is dominant in the CFR and use it for all future testing.				
Instruction	Stopwatch required. Instruct the participant that you will be timing their ability to stand in this position. Have examiner and an assistant stand on each side of participant to assist if participant begins to lose their balance. Use a stopwatch to measure up to 3 attempts.				
Assessment Detail	As soon as the participant is able to exceed 60 seconds (on the first or second trial), no subsequent trials are required and score of zero is recorded for the remaining trial(s).				
Score	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	1 minute or longer	Less than 1 minute, greater than 45 seconds	Less than 45 seconds, greater than 30 seconds	Less than 30 seconds, greater than 15 seconds	Less than 15 seconds or needs hands held by assistant/device, or cannot assume start position.
Notes	Participant can seek support in between trials; holding on to the table, chair or examiner.				

E. UPRIGHT STABILITY					
5. Stance on Dominant Foot					
Position	Participant achieves single limb stance by flexing hip of non-dominant leg and keeping knee in an extended position. Heel should be several inches off the floor.				
Instruction	Stopwatch required. Instruct the participant that you will be timing their ability to stand on one foot. With examiner in front of participant, have an assistant stand behind or on side of participant to assist if participant begins to sway. Use a stopwatch to measure up to 3 attempts.				
Assessment Detail	As soon as the participant is able to exceed 60 seconds (on the first or second trial), no subsequent trials are required and score of zero is recorded for the remaining trial(s).				
Score	0.0	1.0	2.0	3.0	4.0
Scoring Specifics	1 minute or longer	Less than 1 minute, greater than 45 seconds	Less than 45 seconds, greater than 30 seconds	Less than 30 seconds, greater than 15 seconds	Less than 15 seconds or needs hands held by assistant/device, or cannot assume start position.
Notes	Participant can seek support in between trials; holding on to the table, chair or examiner.				

E. UPRIGHT STABILITY					
6. Tandem Walk					
Position	Participant will tandem walk at least 8 steps in a straight line. Performed in hallway or long room with no furniture within reach of 1m/3ft and no loose carpet. Demonstrate task for the participant, specifying how the heel and toe touch with each step. The heel of the front foot should touch the toes of the foot in back as they take steps. Arms can be extended for more balance. Always do this test on the same surface/location (bare floor).				
Instruction	Instruct participant to begin in a tandem stance and then to begin walk with at least 8 steps. Examiner and assistant can walk close to participant for safety if needed.				
Assessment Detail	Count the number of steps completed in a single trial. Score based on number of steps.				
Score	0.0	1.0	2.0	3.0	
Scoring Specifics	Normal (able to tandem walk 8 or greater sequential steps)	Able to tandem walk greater than 4 sequential steps, but less than 8	Able to tandem walk 4 or less steps before losing balance	Too poorly coordinated to attempt task	
Notes					

E. UPRIGHT STABILITY						
7. Gait						
Position	Standing with or without an assistive walking device. Performed in hallway or long room with no furniture within reach of 1m/3ft and no loose carpet.					
Instruction	Observe participant walking at a typical pace in one direction for 8 meters or 25 feet participant					
Assessment Detail	Scoring based on amount of ataxia and if assistive devices are required.					
Score	0.0	1.0	2.0	3.0	4.0	5.0
Scoring Specifics	Normal	Mild ataxia / veering / difficulty in turning; no cane/other support needed to be safe	Walks with definite ataxia; may need intermittent support/examiner needs to walk with participant for safety	Moderate ataxia / veering / difficulty turning; walking requires cane/holding onto examiner with one hand for safety	Severe ataxia / veering; walker or both hands of examiner needed	Cannot walk even with assistance (wheelchair bound)
Notes						

--- END of mFARS ---

Friedreich's Ataxia Rating Scale (FARS)

The following items are assessed, in addition to the above, only if performing the full FARS.

A. BULBAR							
1. Facial Atrophy, Fasciculation, Action Myoclonus, And Weakness							
Instruction	Inspect and activate facial muscles; Can you smile for me?						
Assessment Detail	Most participants with FA do not have significant facial atrophy. If mild facial atrophy is noted, score as per score specifics. Increments of 0.5 may be used if examiner feels an item falls between two severities. Note, If the participant has only a transverse smile, score is 0.5.						
Score	0.0	0.5	1.0	1.5	2.0	2.5	3.0
Scoring Specifics	None		Fasciculations or action myoclonus, but no atrophy		Atrophy present but not profound or complete		Profound atrophy and weakness

A. BULBAR							
2. Tongue Atrophy, Fasciculation, Action Myoclonus, And Weakness							
Instruction	Inspect tongue; Can you stick out your tongue?						
Assessment Detail	Most participants with FA do not have significant tongue atrophy. If mild tongue atrophy is noted, score as per score specifics. Increments of 0.5 may be used if examiner feels an item falls between two severities.						
Score	0.0	0.5	1.0	1.5	2.0	2.5	3.0
Scoring Specifics	Normal		Fasciculations or action myoclonus, but no atrophy		Atrophy present but not profound or complete		Profound atrophy and weakness

D. Peripheral Nervous System, Items D 1 – 5

Prior to transferring participant to a seated position at the edge of the mat table for items in section D, make sure that the participant has at least gravity-eliminated strength in #2 below. Use testing positions on the mat table's surface as needed.

D. PERIPHERAL NERVOUS SYSTEM			
1. Muscle Atrophy			
Position	The participant should sit with 90 degrees of hip flexion, 90 degrees of knee flexion, and feet supported. If a participant cannot sit unsupported in the defined chair, they may stay seated in their adapted seating system.		
Instruction	If atrophy is present or severe, indicate location of atrophy		
Assessment Detail	Score most severe atrophy in either upper or lower limb. Atrophy is scored from denervation and not from lack of use. <i>Score is provided for each side of the body; right and left.</i>		
Score	0.0	1.0	2.0
Scoring Specifics	None	Present Mild/moderate	Severe Total wasting

D. PERIPHERAL NERVOUS SYSTEM

2. Muscle Weakness

Position	The participant should sit with 90 degrees of hip flexion, 90 degrees of knee flexion, and feet supported. If a participant cannot sit unsupported in the defined chair, they may stay seated in their adapted seating system.					
Instruction	Test deltoids, interossei, iliopsoas, and tibial anterior.					
Assessment Detail	Score most severe weakness in either upper or lower limb. <i>Score is provided for each side of the body; right and left.</i>					
Score	0.0	1.0	2.0	3.0	4.0	5.0
Scoring Specifics	None (5/5)	Mild (movement against resistance but not full power 4/5)	Moderate (movement against gravity but not with added resistance 3/5)	Severe (movement of joint but not against gravity 2/5)	Near paralysis (muscular activity without movement 1/5)	Total paralysis (0/5)

D. PERIPHERAL NERVOUS SYSTEM

3. Vibratory Sense

Position	The participant should sit with 90 degrees of hip flexion, 90 degrees of knee flexion, and feet supported. If participant has the hamstring flexibility their legs may be raised using the elevated leg rests. If a participant cannot sit unsupported in the defined chair, they may stay seated in their adapted seating system.		
Instruction	Hands should be placed on their thighs and feet should be comfortably positioned on a support surface. The examiner's hand may function as a support surface under the toe being tested. Educate participant regarding the sensation they will experience at their digit. Use a 128-cps tuning fork set to full vibration. Ask participant to close their eyes. Examiner supports the ventral side distal phalanx and strikes the tuning fork. Start timer as examiner places the tuning fork over the dorsal aspect of the distal phalanx (of great toe or index finger). Use the phrase, "Tell me when the buzzing or vibration goes away".		
Assessment Detail	Vibration sense is recorded (in seconds to the 10 th decimal place) and then given a score depending on the extent of impairment. Abnormal is less than 15 seconds for toes and less than 25 seconds for hands. <i>Score is provided for each side of the body; right and left.</i>		
Score	0.0	1.0	2.0
Scoring Specifics	Normal	Impaired at toes or fingers	Impaired at both toes and fingers

D. PERIPHERAL NERVOUS SYSTEM

4. Positional Sense

Position	The participant should sit with 90 degrees of hip flexion, 90 degrees of knee flexion, and feet supported on floor or raised surface. If a participant cannot sit unsupported in the defined chair, they may stay seated in their adapted seating system.		
Instruction	Test 5 times each using a minimal random movement of 3-4 degrees of the distal interphalangeal joints of index finger and big toe. Make these minimal movements holding the lateral sides of joint with one hand and the distal end with the other hand. Begin with joint in a neutral position, and move to either direction and hold position till participant replies answer. After the participant replies, return to the neutral position before performing next movement. State “Which way did I move it from where it was before?” and instruct participants to answer “up”, “down”, or “I don’t know” after each movement.		
Assessment Detail	Impaired with two or more incorrect attempts. Allow one mistake to acknowledge participant focus. Participant may get 4/5 correct and score a 0. <i>Score is provided for each side of the body; right and left.</i>		
Score	0.0	1.0	2.0
Scoring Specifics	Normal	Impaired at toes or fingers	Impaired at both toes and fingers

D. PERIPHERAL NERVOUS SYSTEM

5. Deep Tendon Reflexes (DTR)

Position	The participant should sit with 90 degrees of hip flexion, 90 degrees of knee flexion, and feet supported on floor or raised surface. If a participant cannot sit unsupported in the defined chair, they may stay seated in their adapted seating system.		
Instruction	This test is measuring whether or not the subject has normal reflexes. Examiner will tap the subject on both the left and right the elbow (BJ), wrist (BrJ), knee (KJ), and ankle (AJ), to test whether they have a normal reflex response to this. Record response for each location. Scores of reflexes are measured independently and range from 0 to 4. Then upper and lower extremity are measured by side separately (i.e. right upper extremity, right lower extremity, left upper extremity, left lower extremity) and the total score for the right side (combination arm and leg) and the left side (combination arm and leg) are tabulated separately based on the scores of the individual joints. Generally, subjects will have absent reflexes (0) and their overall score for DTR will be 2 for each side. See scoring table below.		
Assessment Detail	Score is applied for overall assessment across the 4 locations on each side of the body. 0 = absent, 1 = hyporeflexia, 2 = normal, 3 = hyperreflexia, 4 = pathologic hyperreflexia		
Score	0.0	1.0	2.0
Scoring Specifics	All reflexes are graded normal (either a 1 or a 2 for all reflexes)	Either areflexia (score 0) or hyperreflexia (score 3 or 4) in only upper or only lower limbs	Either areflexia or hyperreflexia in both upper and lower limbs

Calculating the final reflex scores can be difficult if patients have a mixture of areflexia, hyporeflexia, normal, etc. reflexes. When calculating the total DTR: Normal over rules abnormal - so if the patient has a normal value for one extremity (i.e. a 1 or 2) and an abnormal for the other (i.e. a 0, 3 or 4) the limb is still scored as normal. Use **table 1** to refer to examples of different combinations of reflex scores to help calculate final right or left side scores. Please also confirm that these scores are correct with examiner before data entry.

Table 1:

Limb	Right	Left
BJ AND BrJ	0 OR 3 OR 4	0 OR 3 OR 4
KJ AND AJ	0 OR 3 OR 4	0 OR 3 OR 4
Total DTR	2	2
Limb	Right	Left
BJ and/or BrJ	1 or 2	1 or 2
KJ and/or AJ	1 or 2	1 or 2
Total DTR	0	0
Limb	Right	Left
BJ and/or BrJ	0 and 1	0 and 1
KJ and/or AJ	2 and 3	1 or 2
Total DTR	0	0
Limb	Right	Left
BJ and/or BrJ	2 or 3	1 or 2
KJ and AJ	0 and 1 or 0 and 2	0 and 1 or 0 and 2
Total DTR	0	0
Limb	Right	Left
BJ and/or BrJ	0 and 1	0 and 1
KJ and/or AJ	1 or 2	2 and 3
Total DTR	0	0
Limb	Right	Left
BJ and/or BrJ	1	2 and 3
KJ and/or AJ	0 and 1	0 and 1
Total DTR	0	0
Limb	Right	Left
BJ and/or BrJ	2 and 3	0 and 1
KJ and/or AJ	1 or 2	2 and 3
Total DTR	0	0
Limb	Right	Left
BJ and/or BrJ	0 OR 3 OR 4	0 OR 3 OR 4
KJ AND AJ	1 or 2	1 or 2
Total DTR	1	1
Limb	Right	Left
BJ and/or BrJ	1 or 2	1 or 2
KJ AND AJ	0 OR 3 OR 4	0 OR 3 OR 4
Total DTR	1	1
Limb	Right	Left
BJ AND BrJ	0 OR 3 OR 4	0 OR 3 OR 4
KJ and/or AJ	1 or 2	1 or 2
Total DTR	1	1

0= No areflexia

1= Areflexia or mild hyperreflexia in either upper or lower limbs

2= Generalized areflexia or pathologic hyperreflexia

References

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2. Tai G, Corben LA, Gurrin L, Yiu EM, Churchyard A, Fahey M, Hoare B, Downie S, Delatycki MB. A study of up to 12 years of follow-up of Friedreich ataxia utilising four measurement tools. *Journal of neurology, neurosurgery, and psychiatry*. 2014. Epub 2014/08/13. doi: 10.1136/jnnp-2014-308022. PubMed PMID: 25112308.
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