



Disease severity index - Autosomal recessive spastic ataxia of Charlevoix-Saguenay

DSI-ARSACS[©]

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1. Administration

The following equipment is required to administer the scale:

- Short guide to administering the scale
- Scale scoring sheet (see p. 2-3)
- Chair with back and armrests
- Pillow or cushion
- Standard size pen
- Stopwatch
- 2 circles 6 cm in diameter, 1 red circle 2 cm in diameter
- Velcro band of 30 cm
- 8-metre walking distance
- Walking belt
- 2 yellow lines of 30 cm, parallels, 30 cm apart
- 128 Hz tuning fork
- Examination bed

Client's clothing and equipment

The subject should wear loose, comfortable clothing and suitable shoes for the assessment. He should also wear his orthotics and glasses and bring his walking aid, if he uses one.

Timed items: The stopwatch icon beside the title of an item indicates that the test is timed.



Demonstration of items: To ensure the subject fully understands how to perform the test, the examiner should always demonstrate the test when instructing the subject.

Standardized starting position: It is important to use the standardized starting position described for each item.

Item scoring: If in doubt, enter the highest score (ex : 3→4). When applicable, record the continuous value first, and then attribute a score according to response options in the scoring sheet.

Client safety: If, in the examiner's judgement, it would not be safe for the subject to perform the item, it should not be assessed. The highest score should be given.

Rest periods: Rest periods are acceptable between items, as required.

Subject's refusal to be assessed for certain items: If the subject refuses because he is unwilling to cooperate, the item should not be scored. However, if he refuses because he feels he is unable to perform the task, he should be given the highest score.

*** Note: The masculine form is used merely to simplify the text.

2. Scoring sheet

DISEASE SEVERITY INDEX FOR AUTOSOMAL RECESSIVE SPASTIC ATAXIA OF CHARLEVOIX-SAGUENAY

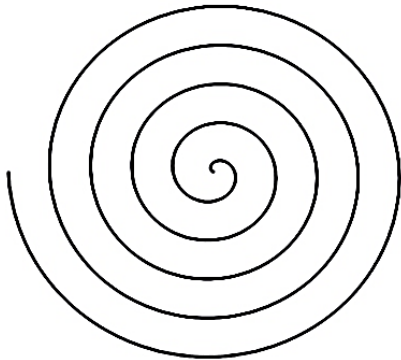
Name:	General condition and remarks
Age:	
Date/time:	
ID number:	Level of participation during the test
Dominance: <input type="checkbox"/> R <input type="checkbox"/> L	

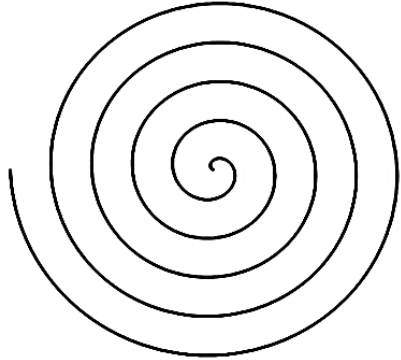
Test	Scoring	Score	
1. Speech during normal conversation	0. Normal 1. Speech slightly impaired (slight anomaly in rhythm and clarity) 2. Speech impaired but easy to understand 3. Occasional words difficult to understand 4. Many words difficult to understand 5. Few words understandable or speech unintelligible	= /5	Language /5
2. Archimedes spiral (see back)	0. Normal (less than 1 cm traced outside the line) 1. Spiral slightly irregular (more than 1 cm traced outside the line), <30 s 2. Spiral slightly irregular (more than 1 cm traced outside the line), ≥30 s 3. Spiral irregular (tracing touches or crosses the adjacent line or another line) 4. Spiral very irregular (tracing touches or crosses two lines or more)	Dominant hand = /5	Upper limbs /9
3. Standardized finger-to-nose test	0. > 20 repetitions 1. 16 to 20 repetitions 2. 11 to 15 repetitions 3. 6 to 10 repetitions 4. ≤ 5 repetitions	# target in 20s: _____ Dominant = /4	
4. Mobility <input type="checkbox"/> Without orthotic(s) <input type="checkbox"/> With orthotic(s)	0. Walks normally 1. Gait abnormal, without aid; walks 8 m in ≤ 10 s 2. Gait abnormal, without aid; walks 8 m in > 10 s and ≤ 20 s 3. Walks with unilateral aid (cane or crutch) and walks 8 m in ≤ 20 s or walks without aid and walks 8 m in > 20 s 4. Walks with bilateral aid (canes, forearm crutches, walker) and walks 8 m in ≤ 20 s or walks with unilateral aid and walks 8 m in > 20 s 5. Walks with bilateral aid and walks 8 m in > 20 s 6. Walking limited to a few steps with bilateral aid, does not exceed 8 m 7. Confined to wheelchair, independent for transfers with or without assistive device(s) 8. Confined to wheelchair, dependent for transfers	= /8	Mobility /8

5. Muscle tone (Modified Ashworth)	0. Normal	Hip adductors	Right /5 Left /5	Mean = /5	Lower limbs /12
	1. Slight ↑ in muscle tone manifested by a catch and release or by minimal resistance at the end of the ROM (1)				
2. Slight ↑ in muscle tone manifested by a catch followed by minimal resistance throughout the remainder (less than half) of the ROM (1+)					
3. More marked ↑ in muscle tone through most of the ROM (more than half), but affected part(s) easily moved (2)					
4. Considerable ↑ in muscle tone, passive movement difficult. (3)					
5. Affected part(s) rigid in flexion or extension, abduction or adduction (4)					
6. Vibration lateral malleolus	0. 12 seconds or more	Seconds Right = Left =	Score Right = /3 Left = /3	Mean = /3	
	1. 8,00-11,99 seconds				
	2. 4,00-7,99 seconds				
	3. Less than 4 seconds				
7. Circle with foot	0. Circle normal (diameter of 30 cm)	Right = /4 Left = /4	Mean = /4		
	1. Circumduction with angles in the completed circle (diameter of 30 cm)				
	2. Significant decomposition of the completed circle (chaotic movements)				
	3. Starts moving the heel (circle incomplete)				
4. Unable to initiate movement					
8. Bladder function	0. No bladder symptoms	= /4	Total score /38	Bladder /4	
	1. Urinary urgency and/or pollakiuria and/or nycturia and/or occasional incontinence, not requiring treatment				
	2. Urinary urgency and/or pollakiuria and/or nycturia and/or occasional incontinence, requiring treatment				
	3. Urinary incontinence, may be controlled with treatment				
4. Urinary incontinence despite treatment					

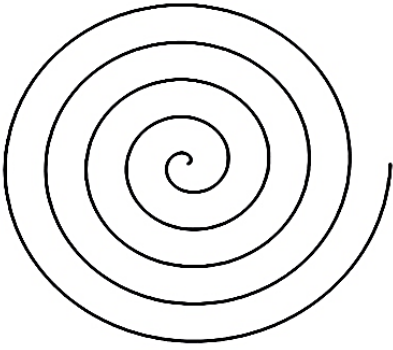
Examiner's signature: _____

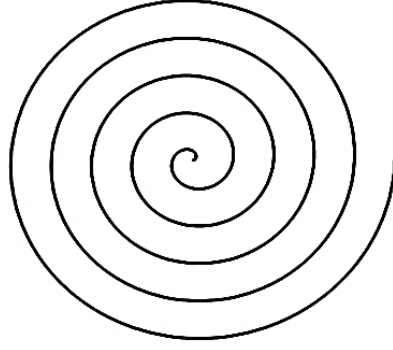
Archimedes spiral





Right-handed





Left-handed

3. Items' description

3.1. Speech

Procedure

Observe during normal conversation throughout the assessment.

Scoring

0. Normal
1. Speech slightly impaired (slight anomaly in rhythm and clarity)
2. Speech impaired but easy to understand
3. Occasional words difficult to understand
4. Many words difficult to understand
5. Few words understandable or speech unintelligible



3.2. Archimedes spiral

Equipment required

Chair
 Table of a suitable height
 Standard size pen
 Spirals on the scoring sheet
 Stopwatch

Position

The subject is seated at the table, with the elbow and forearm supported, the feet flat on the floor. He performs the task with his dominant hand on the appropriate spiral indicated on the scoring sheet. A standard size pen is used. He may position the sheet as he likes before the test and stabilize the sheet with his other hand. The examiner may not stabilize the sheet.

Procedure

The subject performs the test on the two spirals. The examiner scores the best spiral and notes the time taken by the subject to complete each spiral.

Instructions

*Draw a spiral by following the line, **without** lifting the pen. Start at the centre and continue until the end of the line. The quality of the drawing is important, not rapidity.*

Scoring (Score the best of the two)

0. Normal (less than 1 cm traced outside the line)
1. Spiral slightly irregular (more than 1 cm traced outside the line), <30sec
2. Spiral slightly irregular (more than 1 cm traced outside the line), ≥30sec
3. Spiral irregular (tracing touches or crosses the adjacent line or another line)
4. Spiral very irregular (tracing touches or crosses two lines or more)
5. Spiral unrecognizable or incomplete

3.3. Standardized finger-to-nose test



Equipment required

Chair
Stopwatch
1 red circle 2 cm in diameter
Velcro band of 30 cm

Position

The subject is seated on a chair, facing wall, back well supported (a pillow may be placed behind his back), the feet flat on the floor. The subject's chair is positioned so that subject's nose is found at 45 cm straight front wall. The target is a red circle 2 cm in diameter that can be moved on a vertical axis (velcro band) according to the subject's nose height.

Procedure

Using the index finger, the subject must touch, alternately and as quickly as possible in a 20-second period, the end of his nose and a horizontal target 45 cm away. If the subject does not touch the target directly and accurately, he is instructed to move his finger to touch the target before returning to his nose. The subject practices for few repetitions on each side. He performs the test with his dominant hand first and then with his other hand. The execution time is measured with the stopwatch and the number of repetitions (# of touched targets) is recorded. Also note the number of missed targets. Two trials on each side are executed.

Instructions

Put your (right or left) index finger on your nose. At my signal, touch the target and then return to your nose as quickly and accurately as possible during 20 seconds. You may practice and, when you are ready, start at my signal (1,2,3,GO). If you miss a target, I will ask you to return to the target before returning to your nose.

Scoring

0. > 20 repetitions
1. 16 to 20 repetitions
2. 11 to 15 repetitions
3. 6 to 10 repetitions
4. 5 repetitions or less

3.4. Mobility



Equipment required

Distance of 8 meters marked on the floor, identification line 2 metres before the start line

Stopwatch

Walking aid, if any

Position

The subject is wearing his shoes and orthotics and stands 2 metres before the start line with his walking aid.

Procedure

The examiner times how long it takes him to walk 8 metres. The subject's feet must cross the finish line.

Instructions

Walk at your normal pace until I say stop.

Scoring (Note if wearing orthotic(s) or not)

0. Walks normally
1. Gait abnormal, without aid and walks 8 m in ≤ 10 s
2. Gait abnormal, without aid and walks 8 m in ≤ 20 s
3. Walks with unilateral aid (cane or crutch) and walks 8 m in ≤ 20 s or walks without aid and walks 8 m in > 20 s
4. Walks with bilateral aid (canes, forearm crutches, walker) and walks 8 m in ≤ 20 s or walks with unilateral aid and walks 8 m in > 20 s
5. Walks with bilateral aid and walks 8 m in > 20 s. Sometimes uses wheelchair for daily mobility
6. Walking limited to a few steps with bilateral aid, does not exceed 8 m. Mainly uses wheelchair for daily mobility
7. Confined to wheelchair, independent for transfers with or without assistive device(s)
8. Confined to wheelchair, dependent for transfers

3.5. Muscle tone

Equipment required

Chair

Pillow

Position

The subject is seated, back supported (a pillow may be placed behind his back), the feet flat on the floor.

Procedure

The tone of the hip adductors and knee extensors is assessed. Both limbs are assessed, one after the other, except for the hip adductors which are assessed simultaneously. The examiner makes a rapid passive movement through the available range of motion and repeats the movement 3 times.

Instructions

Stay very relaxed while I move your leg.

Hip adductors: Place the subject's knees in a maximally adducted position and move to a position of maximal abduction.

Scoring (Modified Ashworth Scale)¹

0. Normal
1. Slight ↑ in muscle tone manifested by a catch and release or by minimal resistance at the end of the ROM (1)
2. Slight ↑ in muscle tone manifested by a catch followed by minimal resistance throughout the remainder (less than half) of the ROM (1+)
3. More marked ↑ in muscle tone through most of the ROM (more than half), but affected part(s) easily moved (2)
4. Considerable ↑ in muscle tone, passive movement difficult. (3)
5. Affected part(s) rigid in flexion or extension, abduction or adduction. (4)

3.6. Vibration sense**Equipment required**

Chair
Pillow or cushion
Stopwatch
128 Hz tuning fork

Position

The subject is seated, back supported (a pillow may be placed behind his back).

Procedure

Use a 128 Hz tuning fork, at maximum vibration. Demonstrate the vibration on a metacarpophalangeal joint. You must hit to obtain an audible sound in all tests. The test is performed on the lateral malleolus. Repeat the test if the first attempt is not conclusive.

Instructions

Tell me if you feel the vibration and when the vibration stops.

Scoring

0. 12 seconds or more
1. 8.00-11.99 seconds
2. 4.00-7.99 seconds
3. Less than 4 seconds

3.7. Circle on the floor with foot

Equipment required

Chair
Pillow
2 yellow lines of 30 cm, parallels, 30 cm apart

Position

The subject is seated with his feet flat on the floor. Make sure the subject is stable when seated before performing the test. The subject may stabilize himself by putting his arms on the armrests and a pillow may be placed behind his back.

Procedure

The test starts with the knee flexed to 90°. The subject must trace a circle 30 cm in diameter on the floor in an anticlockwise direction. The subject may practice twice. The test is performed with the dominant leg first and then with the other leg.

Instructions

Trace a circle inwards between the two yellow lines, dragging your feet on the floor.

Scoring

0. Circle normal (diameter of 30 cm)
1. Circumduction with angles in the completed circle (diameter of 30 cm)
2. Significant decomposition of the completed circle (chaotic movements)
3. Starts moving the heel (circle incomplete)
4. Unable to initiate movement

3.8. Bladder function

Definitions

Pollakiuria: need to urinate more than 8 times in 24 h.

Nycturia: need to urinate more than once during sleep.

Procedure

Ask the subject about the presence of symptoms of neurogenic bladder listed on the scoring sheet.

Scoring

0. No bladder symptoms
1. Urinary urgency and/or pollakiuria and/or nycturia, and/or occasional incontinence, not requiring treatment
2. Urinary urgency and/or pollakiuria and/or nycturia, and/or occasional incontinence, requiring treatment
3. Urinary incontinence, can be controlled with treatment
4. Urinary incontinence despite treatment

4. Items' origin

Speech

Adaptation of item #4, *Scale for the Assessment and Rating of Ataxia (SARA)*²

Archimedes spiral

Adaptation of item #14, *International Cooperative Ataxia Rating Scale (ICARS)*³

Standardized finger-to-nose test (SFNT)

*Standardized Finger-Nose Test*⁴

Scoring system developed by GRIMN's research team

Mobility

Adaptation of *Hauser Ambulation Index*⁵

Muscle tone

Modified Ashworth scale¹

Choice of muscle group and standardized position made by GRIMN's research team

Vibration sense

Adaptation of items #41 and 42, *Ataxia Clinical Rating Scale*⁶

Circle on the floor with foot

Adaptation of item #10, *Modified International Cooperative Ataxia Rating Scale (MICARS)*⁷

Bladder function

Adaptation of item #13, *Spastic Paraplegia Rating Scale (SPRS)*⁸

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