

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) 5. Spiral unrecognizable or incomplete	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), or partial human assistance 8. Confined to wheelchair, dependent for transfers (with complete human assistance)	= /8	Mobility /8

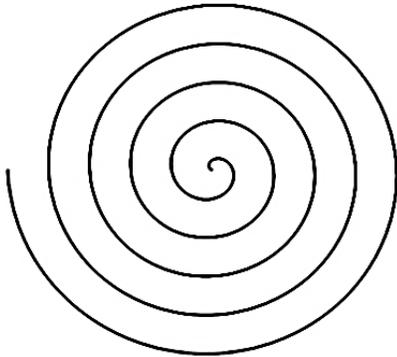
5. Muscle tone (Modified Ashworth)	0. Normal	Hip adductors Right /5 Left /5	Mean = /5
	1. Slight ↑ in muscle tone manifested by a catch and release or by minimal resistance at the end of the ROM (1)		
	2. Slight ↑ in muscle tone manifested by a catch followed by minimal resistance throughout the remainder (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)		
		<i>Seconds</i> Right = Left =	
6. Vibration lateral malleolus	0. 12 seconds or more	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
	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		

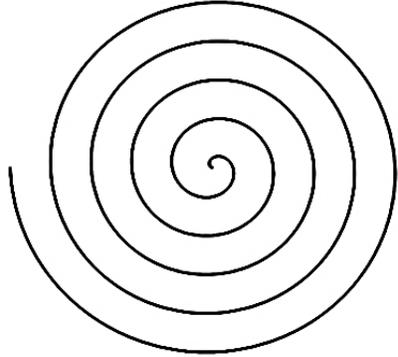
Lower limbs
/12

Bladder
/4

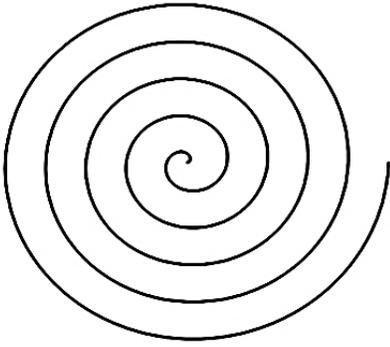
Examiner's signature: _____

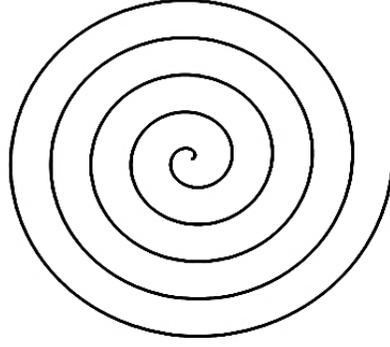
Archimedes spiral





Right-handed





Left-handed