

The ALS Score of Jablecki et al for Disability in Amyotrophic Lateral Sclerosis

Overview: Jablecki et al developed a score for evaluating disability in patients with amyotrophic lateral sclerosis (ALS). This can be used to monitor a patient's decline in ability over time and can be used to estimate survival. The authors are from the University of California and Children's Hospital in San Diego.

Parameters:

- (1) speech
- (2) swallowing
- (3) respiration
- (4) ambulation
- (5) upper extremity strength right
- (6) upper extremity strength left
- (7) upper extremity survey score right
- (8) upper extremity survey score left
- (9) lower extremity strength right
- (10) lower extremity strength left
- (11) lower extremity survey score right
- (12) lower extremity survey score left

Parameter	Finding	Points
speech	normal	0
	mild dysarthria which presents no practical difficulty	1
	moderate dysarthria with speech occasionally difficult to understand	2
	severe dysarthria with speech intelligible to close friend but difficult for casual acquaintance to understand	3
	speech	4
swallowing	normal	0
	mild dysphagia with occasional choking manageable by eating slowly	1
	moderate dysphagia; frequent choking or difficulty swallowing unless certain foods are	2

	avoided	
	severe dysphagia; blenderized diet required	3
	nutrition depends on tube feeding	4
respiration	normal	0
	dyspnea with exertion (walking talking)	1
	dyspnea at rest	2
ambulation	normal	0
	abnormal but walks without assistance	1
	consistently uses a cane or foot brace(s) to walk on own	2
	requires stabilization (walker attendant) to walk	3
	unable to walk and requires assistance to maintain standing balance	4
	unable to maintain standing balance even when assisted	5
	unable to assist with legs during transfers	6
upper extremity survey score right	no cramps or fasciculations normal strength normal rate of finger tapping	0
	cramps or fasciculations present normal strength normal rate of finger tapping	1
	weakness and/or reduced rate of finger tapping with or without cramps or fasciculation	2
upper extremity survey score left	no cramps or fasciculations normal strength normal rate of finger tapping	0
	cramps or fasciculations present normal strength normal rate of finger tapping	1
	weakness and/or reduced rate of finger tapping with or without cramps or fasciculation	2
lower extremity survey score right	no cramps or fasciculations normal strength normal rate of foot tapping	0
	cramps or fasciculations present normal strength normal rate of foot tapping	1
	weakness and/or reduced rate of foot tapping	2

	with or without cramps or fasciculation	
lower extremity survey score left	no cramps or fasciculations normal strength normal rate of foot tapping	0
	cramps or fasciculations present normal strength normal rate of foot tapping	1
	weakness and/or reduced rate of foot tapping with or without cramps or fasciculation	2

Muscle in Upper Extremity Left or Right	Finding	Points
deltoid	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4
biceps	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4
flexor pollicis longus	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4
first dorsal interosseous	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4

Muscle in Lower Extremity Left or Right	Finding	Points
iliopsoas	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4
quadriceps	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4
tibialis anterior	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4
tibialis posterior	normal strength	0
	mild weakness (1 – 25% reduction)	1
	moderate weakness (26 – 50% reduction)	2
	severe weakness (51 – 75% reduction)	3
	useless (76 – 100% reduction)	4

muscle strength score for a limb = (SUM(points for all 4 muscles)) / 4

ALS score = SUM(points for all 12 parameters)

Interpretation:

- minimum score: 0
- maximum score: 40
- The higher the score the greater the disability.
- The score can be used to predict survival time when combine with the patient's age and duration of weakness as discussed in the chapter on neurology.

References:

Jablecki CK Berry C Leach J. Survival prediction in amyotrophic lateral sclerosis. Muscle & Nerve. 1989; 12: 833-841