

Patient-ID

Date of visit

 / / **(O) Onset region**

- O1) **head onset** with dysarthria or dysphagia, also named "bulbar onset"
- O2d) **distal arm onset** with weakness or slowed, poorly coordinated voluntary movements at the distal arm (hand)
- O2p) **proximal arm onset** with weakness at proximal arm (shoulder)
- O3r) **trunk respiratory onset** with weakness at the trunk with hypoventilation
- O3a) **trunk axial onset** with weakness of trunk muscles without concurrent hypoventilation
- O4d) **distal leg onset** with weakness or slowed, poorly coordinated voluntary movements at the distal leg (foot)
- O4p) **proximal leg onset** with weakness or slowed, poorly coordinated voluntary movements at the proximal leg (hip and thigh muscles)

(P) Propagation ("spreading")**Yes, within 12 months**

- PE) **Earlier propagation** Propagation of slowed, poorly coordinated voluntary movements or weakness from the region of onset to another vertically distant body region **within 12 months of symptom onset**.

No, later than 12 months of symptom onset

- PL) **Later propagation** Propagation of slowed, poorly coordinated voluntary movements or weakness from the region of onset to another, vertically distant body region **later than 12 months** of symptom onset. It includes O1, PL (also named „progressive bulbar paralysis, PBP“), O2d/p, PL (also named „flail arm-syndrome“), and O4d/p, PL (also named „flail leg-syndrome.“)

No, onset is less than 12 months.

- PN) **Propagation not classifiable** Propagation of slowed, poorly coordinated voluntary movements or weakness from the region of onset to another, vertically distant body region not yet classifiable as time since symptom **onset is less than 12 months**.

(M) Motor neuron dysfunction

- M0) **balanced motor neuron dysfunction** Balanced combined symptoms of the upper motor neuron (slowed, poorly coordinated voluntary movements, increased reflexes and/or spasticity) and the lower motor neuron (weakness and associated atrophy), also named "classic ALS"
- M1d) **dominant upper motor neuron (UMN) dysfunction** Dominant symptoms of the upper motor neuron (slowed, poorly coordinated voluntary movements, increased reflexes and/or spasticity) and only discrete symptoms of the lower motor neuron (weakness and associated atrophy)
- M1p) **pure UMN dysfunction** Pure symptoms of the upper motor neuron (slowed, poorly coordinated voluntary movements, increased reflexes and/or spasticity) and no evidence of lower motor neuron symptoms (weakness and associated atrophy); also named "primary lateral sclerosis (PLS)" after a disease duration of 48 months
- M2d) **dominant lower motor neuron (LMN) dysfunction** Dominant symptoms of the lower motor neuron (weakness and associated atrophy) and only discrete symptoms of the upper motor neuron (slowed, poorly coordinated voluntary movements, increased reflexes and/or spasticity)
- M2p) **pure LMN dysfunction** Pure symptoms of the lower motor neuron (weakness and associated atrophy) and no evidence of upper motor neuron symptoms (slowed, poorly coordinated voluntary movements, increased reflexes and/or spasticity); also named "progressive muscle atrophy (PMA)" after a disease duration of 48 months
- M3) **dissociated motor neuron dysfunction** Dominant symptoms of the lower motor neuron (weakness and associated atrophy) in the arms and dominant symptoms of the upper motor neuron (slowed, poorly coordinated voluntary movements, increased reflexes and/or spasticity) in the legs