Supplementary Table 2. Summary of excitability changes of peripheral nerves in ALS patients. Abbreviations: asymp., asymptomatic; CMAP, compound muscle action potential; EMG, electromyography; fALS, familial ALS; FPs, fasciculation potentials; H'H ratio, H' response to the maximum H-reflex response; MUs, motor units; n.s., not specified; MUAP, muscle unit action potentials; NCS, nerve conduction studies; SOD1, superoxide dismutase 1; sALS, sporadic ALS; TEd, depolarizing threshold electrotonus; TEh, hyperpolarizing threshold electrotonus; τ_{SD}, strength-duration constant.

region	ALS type	age (average)	method	finding	references
Peripheral nerves	sALS	51 years	NCS in median, sural and peroneal nerves; EMG of interosseus, biceps brachii, vastus lateralis and tibialis anterior muscles	hyperexcitability: FP ↑, double discharging MUs ↑	(Kostera-Pruszczyk et al., 2002)
	sALS	63 years	NCS of median nerve and CMAP recordings of abductor pollicis brevis threshold tracking and current-threshold measurements of median nerve	CMAP ↓ axonal hyperexcitability: τ_{SD} ↑ (CMAP > 5 mV group), TEd ↑, supernormality ↑	(Kanai et al., 2006)
		59.6 years		CMAP ↓ axonal hyperexcitability: τ_{SD} ↑, TEd ↑, TEh ↑ superexcitability ↑	(Vucic and Kiernan, 2006)
	fALS	58 years		CMAP ↓ axonal hyperexcitability: τ_{SD} ↑	(Vucic and Kiernan, 2009)
	sALS	59 years			
	fALS (asymp. SOD1)	40 years		axonal excitability ↔, τ_{SD} ↔	
	n.s.	59.6 years	threshold tracking measurements of ulnar nerve	axonal hyperexcitability (7 patients): TEd ↑ axonal hypoexcitability (4 patients): TEd ↓	(Bostock et al., 1995)
		n.s.	threshold tracking measurements of median nerve	axonal hyperexcitability: TEd ↑, TEh ↑	(Horn et al., 1996)
		59.1 years	NCS of median nerve and CMAP recordings of abductor pollicis brevis; threshold tracking of median nerve	CMAP ↓ axonal hyperexcitability: τ_{SD} ↑, rheobase ↓	(Mogyoros et al., 1998)
		64 years	NCS of median nerve and CMAP recordings of abductor pollicis brevis	CMAP ↓, conduction velocity ↓	(Tamura et al., 2006)
			latent addition with threshold tracking of the median nerve	axonal hyperexcitability: nodal persistent Na^+ conductance ↑, TEd ↑	
		66 years	NCS of median nerve and CMAP recordings of abductor pollicis brevis; threshold tracking of median nerve at wrist (nerve trunk) and at abductor pollicis brevis (distal axons)	CMAP ↓ axonal hyperexcitability: TEd ↑, TEd ↑ (distal axons) > TEd ↑ (nerve trunk)	(Nakata et al., 2006)
		54.7 years	single MUAP recordings of the brachial biceps	hyperexcitability: double discharge firing MUs ↑	(Piotrkiewicz et al., 2008)
		64.0 years	EMG of tibialis anterior	hyperexcitability: complex FPs ↑	(de Carvalho and Swash, 2013)
		46.3 years	soleus H-reflex measurement with EMG	recurrent inhibition ↓, H'/H ratio ↓	(Raynor and Shefner, 1994)
	Riluzole treated ALS patients	54 years	threshold tracking and single MUAP recordings of the abductor pollicis brevis and abductor digiti minimi	CMAP ↓ axonal hypoexcitability: rheobase ↑, threshold for single MU ↑, axonal hyperexcitability: TEd ↑, superexcitability ↑	(Howells et al., 2018)

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