

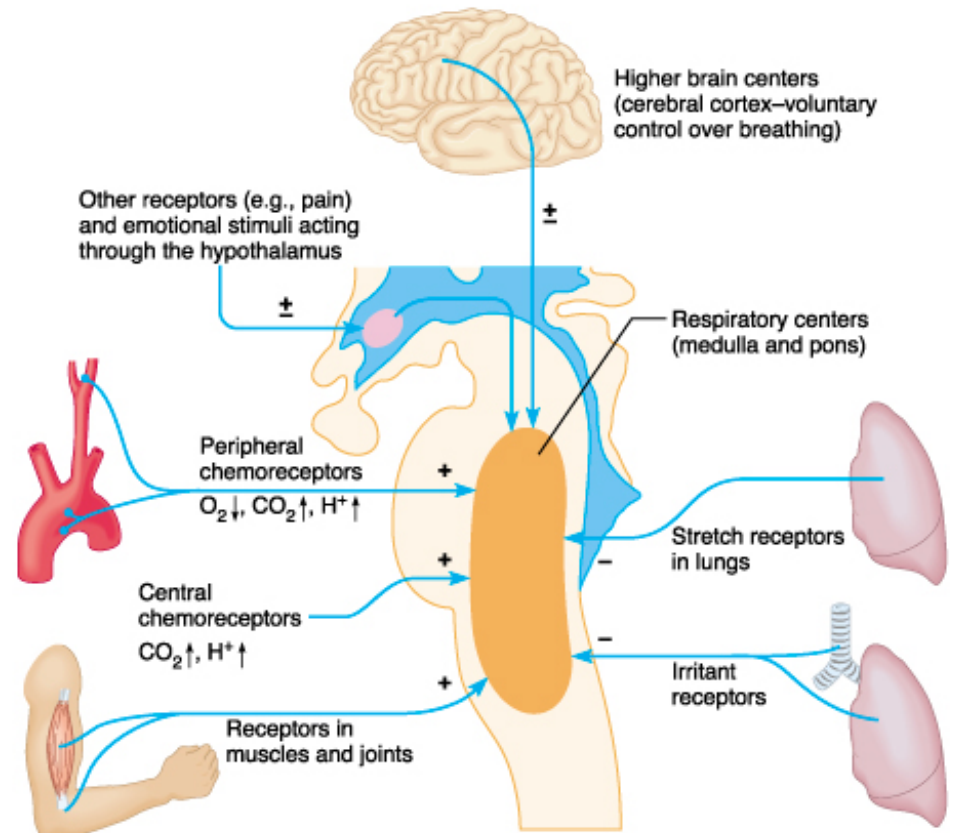
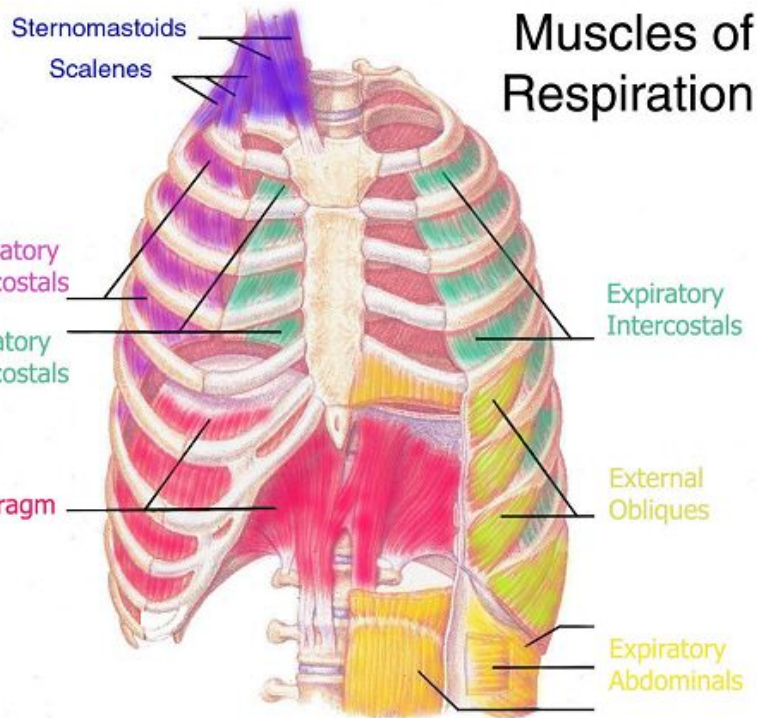


Impact of upper motor neuron degeneration on respiration in wakefulness and sleep in amyotrophic lateral sclerosis (ALS)

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Breathing: complex and vital



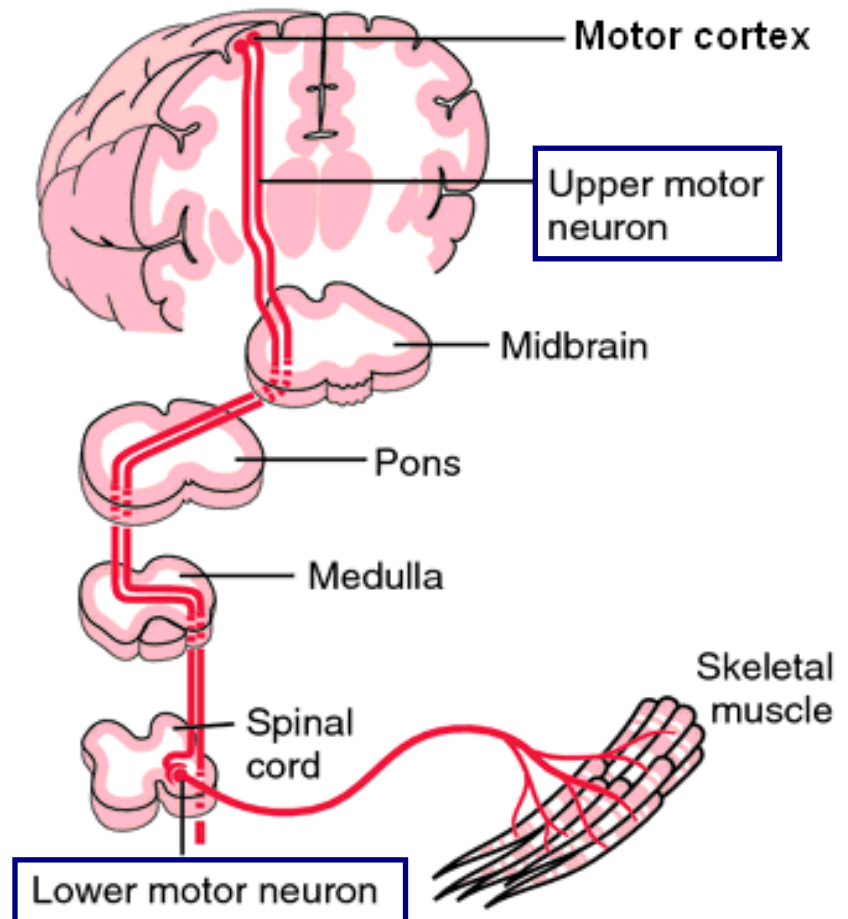
Amyotrophic Lateral Sclerosis (ALS)

- Neurodegenerative disease
- incidence: 6 to 8 / 100.000, ~40 million Europeans
- Marked differences among patients.



Amyotrophic Lateral Sclerosis (ALS)

- Upper and lower motor neurons die out
- Differences among patients
- Lower MN – respiratory muscle weakness
- Upper MN – reduced volitional control over respiratory muscles



Methods: Outcome measures



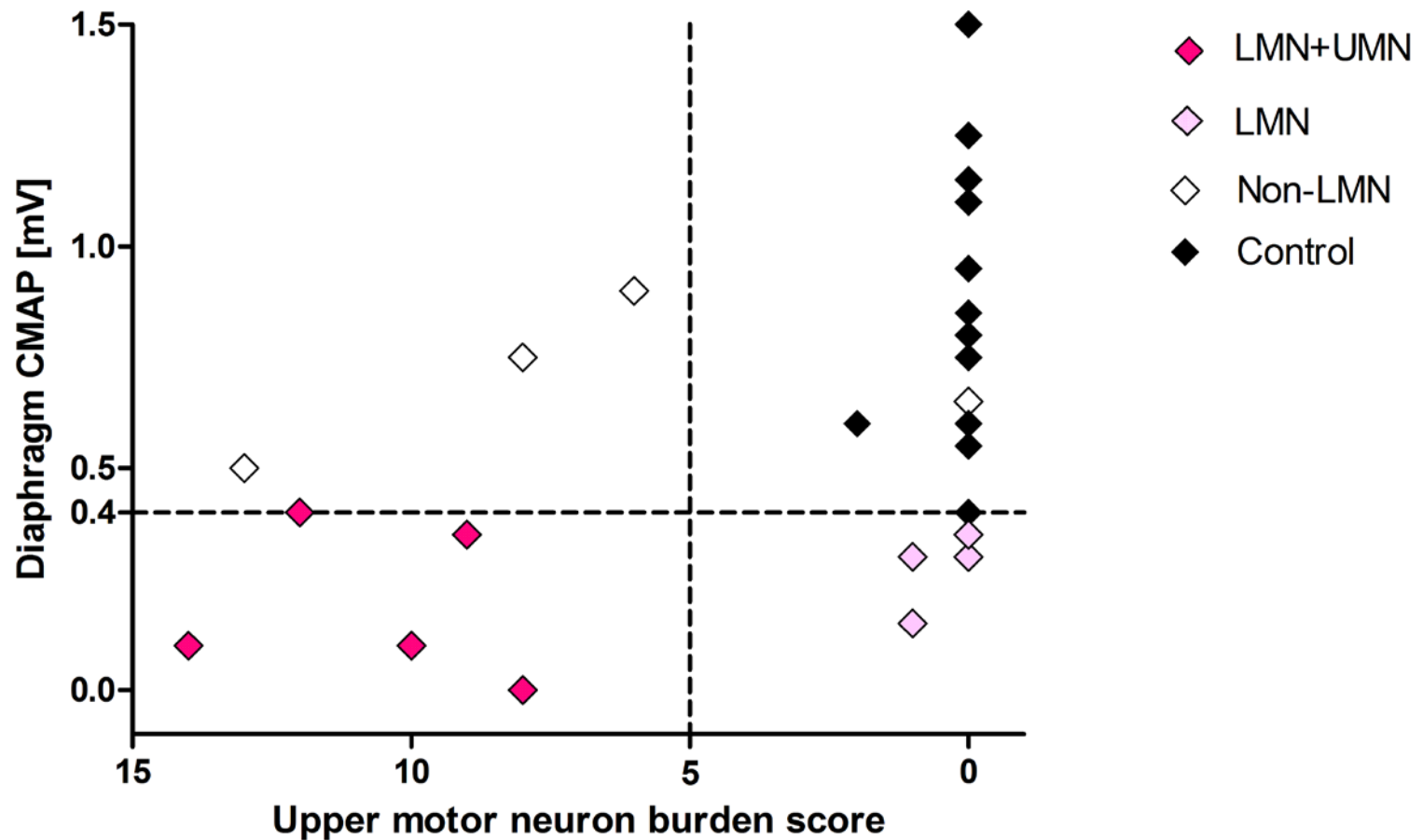


Results: Patient characteristics

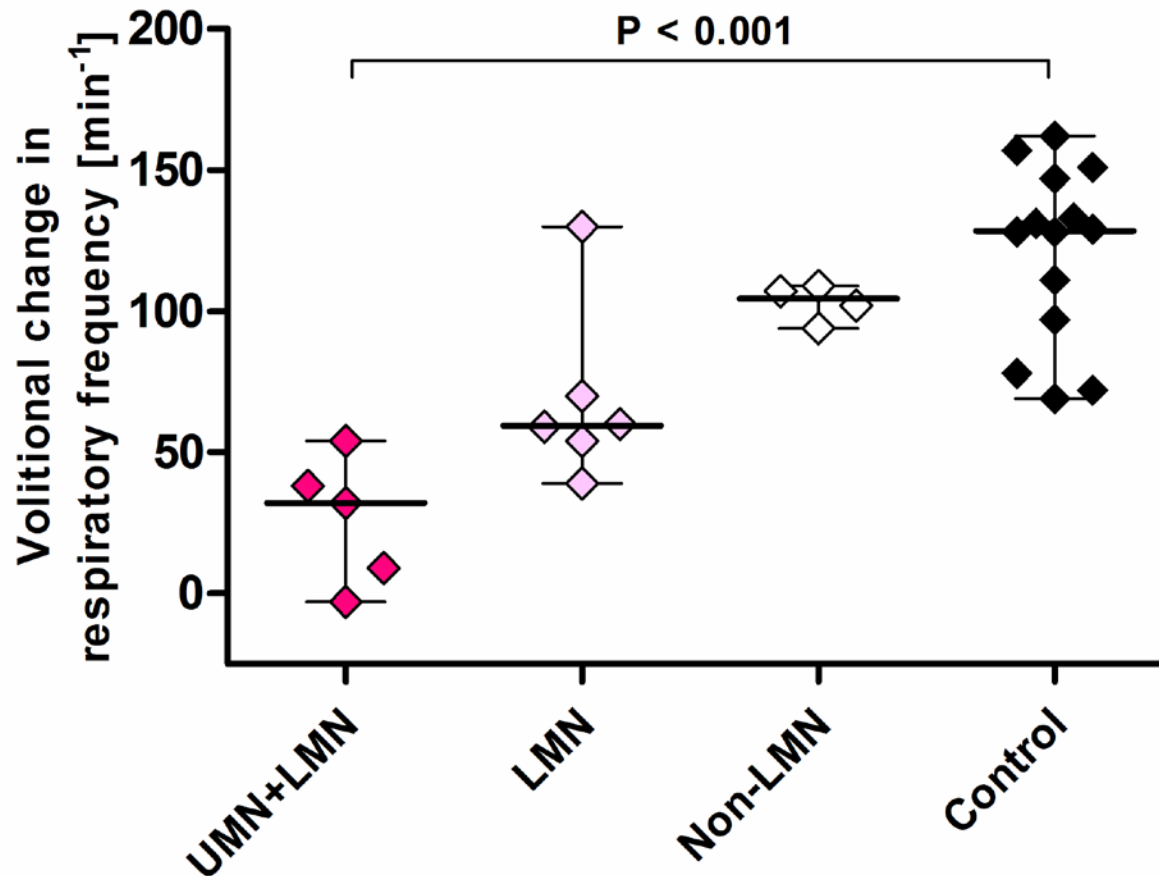
	ALS	Control
Number (females)	15 (7)	14 (6)
Age [years]	60 (9)	60 (9)
BMI [kg/m ²]	23.8 (3.6)	24.1 (3.1)
VC [% pred.]	51 (29, 97) ^{***}	111 (96, 163)
SNIP [kPa]	40 (33) ^{***}	95 (27)



Results: Grouping of patients

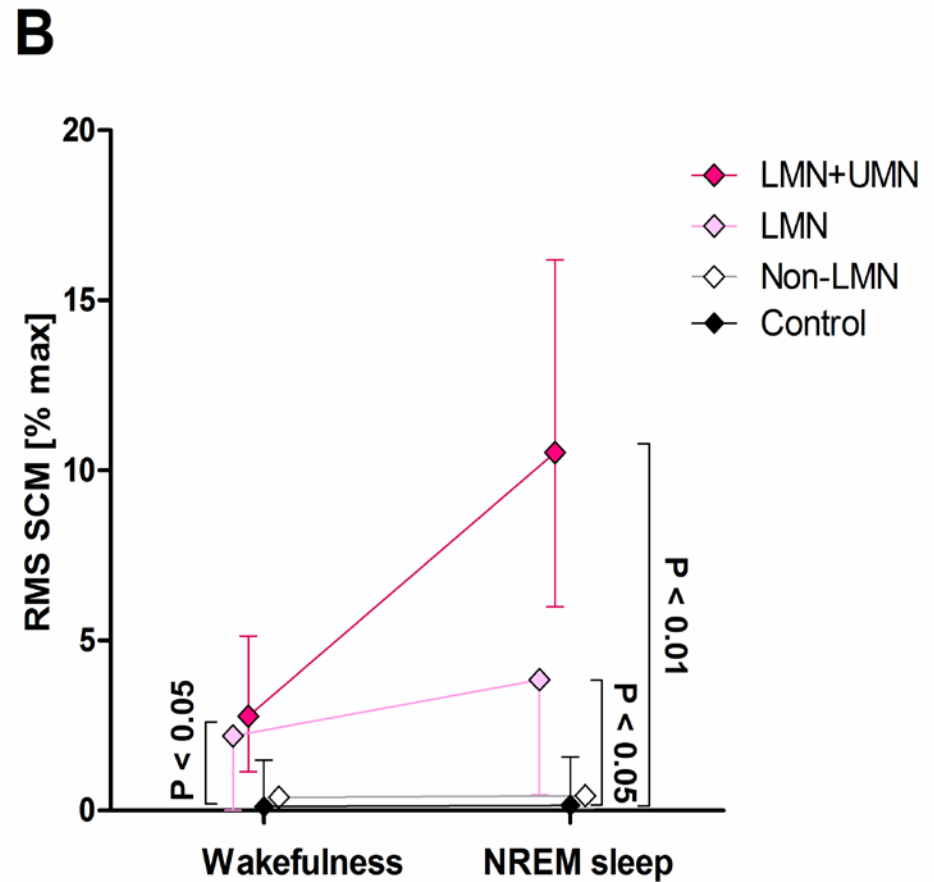
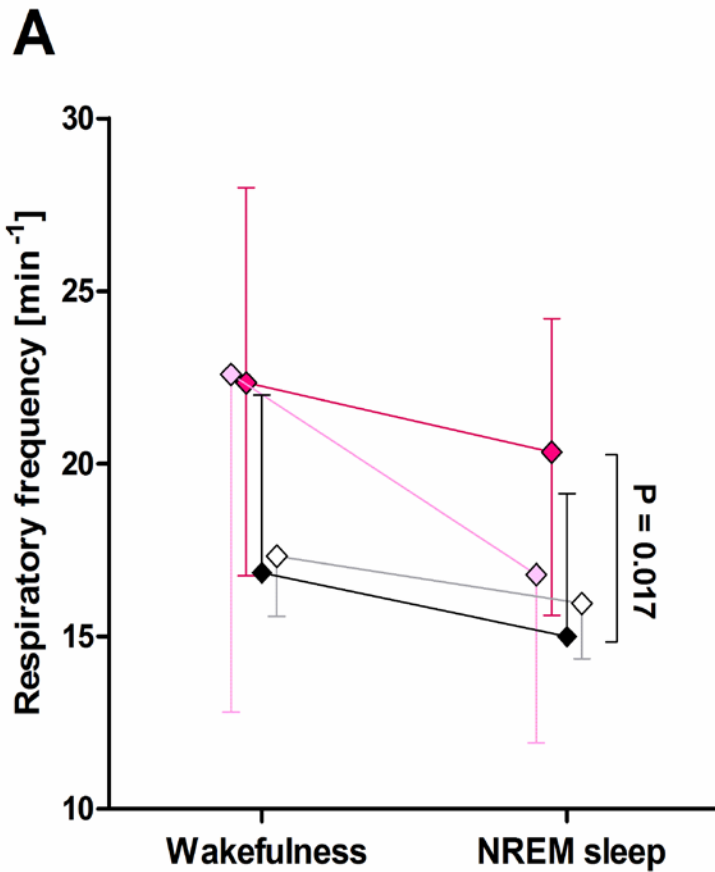


Results: Volitional control of breathing





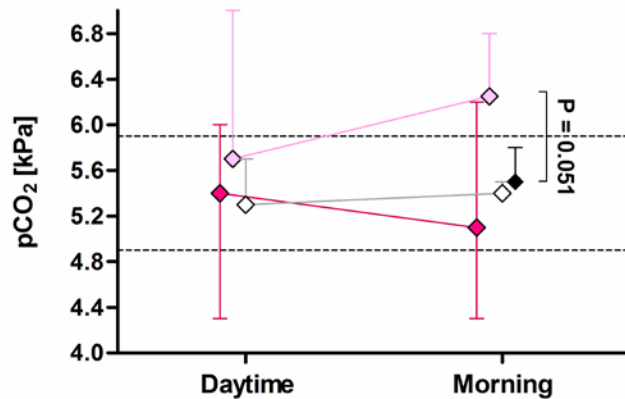
Results: Respiratory drive



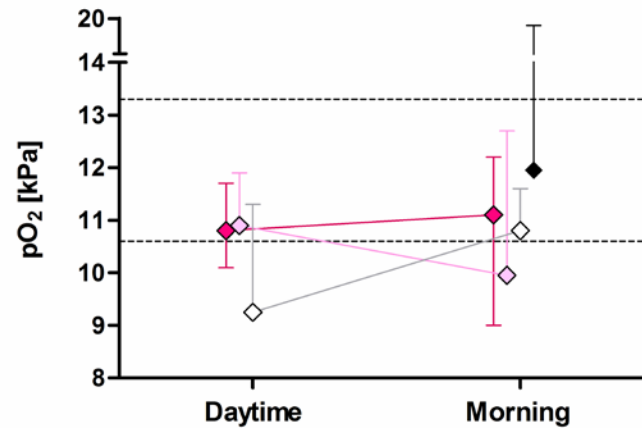


Results: Blood gas analysis

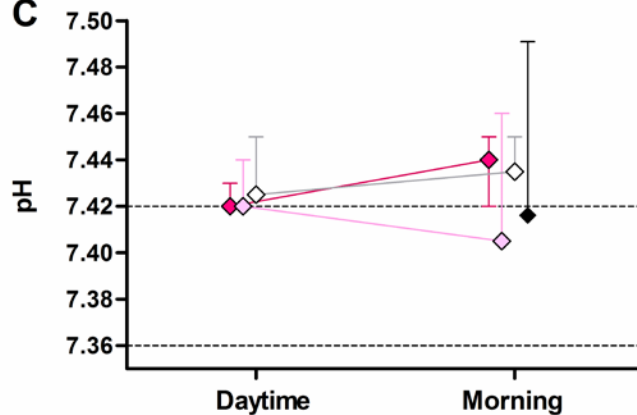
A



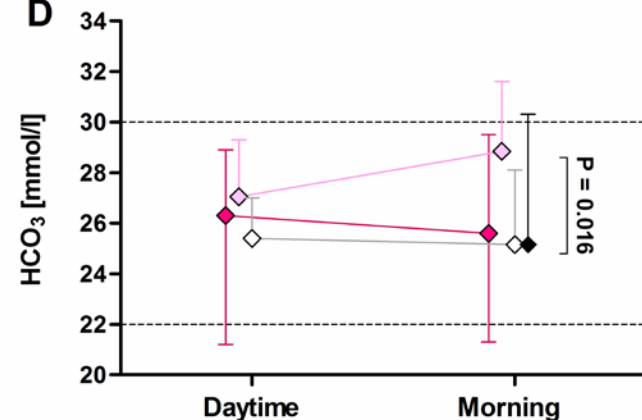
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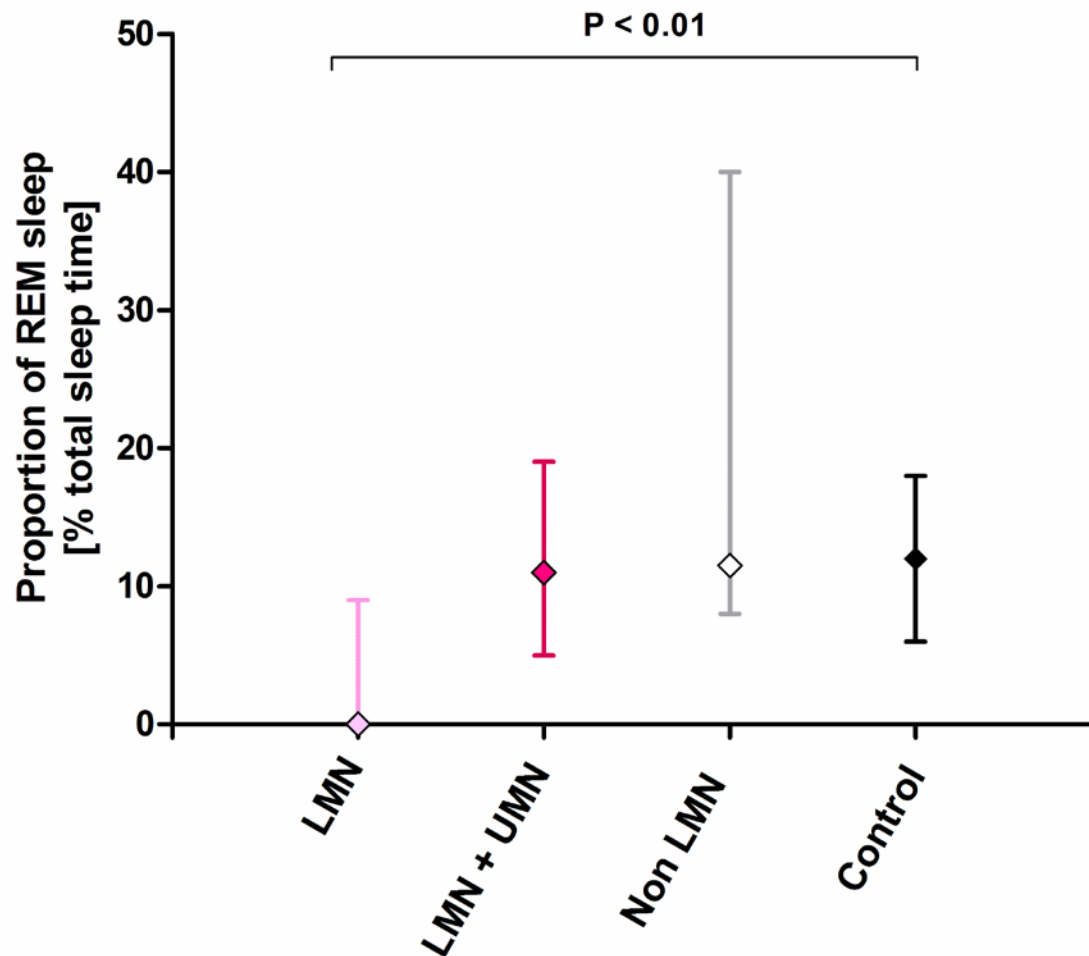


D



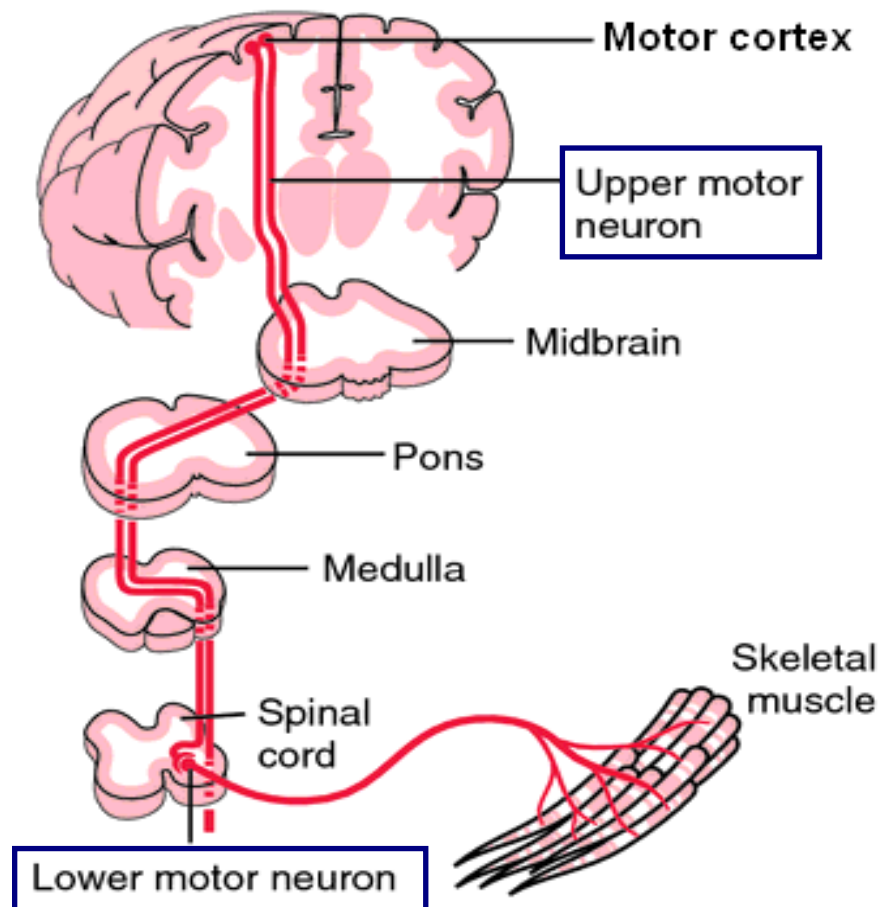


Results: Proportion of REM sleep



Main puzzle:

Why do patients with a more widespread degeneration breathe better?



Answer: Acclimatization



O.K. but ...

Why do not all ALS patients with weak respiratory muscles acclimatize?





Because ...

Healthy people
&
ALS with lower MN
degeneration



	Awake	Asleep
Voluntary breathing	✓	✗
Involuntary breathing	✓	✓
Wakefulness drive to breathe	✓	✗



Because ...

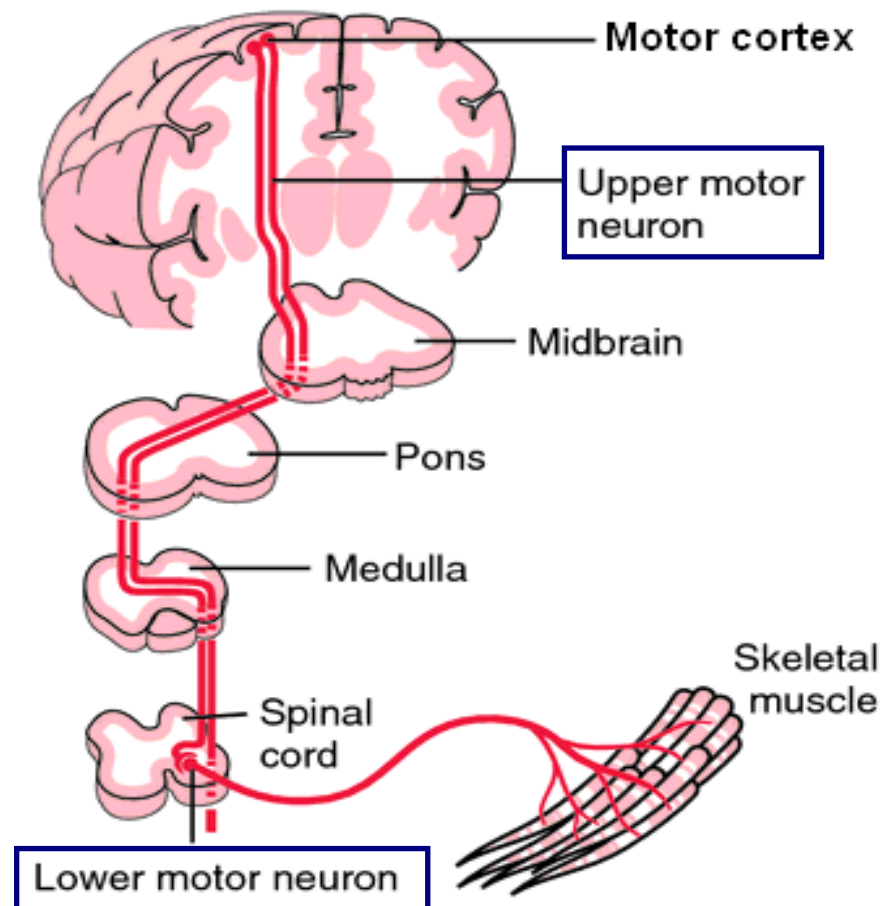


ALS with lower &
upper MN
degeneration

	Awake	Asleep
Voluntary breathing	✓	✗
Involuntary breathing	✓	✓
Wakefulness drive to breathe	?	✗

Take home message

- Carefully observe respiration in ALS and other neurological patients
- If upper and lower motor neurons are injured, acclimatization is possible
- Initial benefit → risk of sudden respiratory failure





Thank you!

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The ALS team

Patients and volunteers

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