Since January 2020 Elsevier has created a COVID-19 resource centre with free information in English and Mandarin on the novel coronavirus COVID-19. The COVID-19 resource centre is hosted on Elsevier Connect, the company's public news and information website.

had a profile of amnesic mild cognitive impairment and 5 (1 VUS) had cognitively-normal profile. At follow up, three ALSbi patients (one with VUS) developed frank frontotemporal dementia (FTD) and one ALSbi developed cognitive impairment (ALScbi). The most common behavioural changes were apathy, mental rigidity and irritability.

Conclusions

Our findings demonstrate that SOD1 patients have early behavioural impairment more commonly than previously reported, which might increase the risk to develop FTD.

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117701

Amyotrophic lateral sclerosis patients' quality of life and their caregiver burden during COVID-19 pandemic

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Background and aims

The two-month lockdown period during COVID-19 pandemic had a general impact on health treatments and care assistance. We wanted to assess Quality of life (QoL) of ALS patients and the burden of their caregivers during that period.

Methods

60 patients and 59 caregivers, visited in telemedicine during March 2020, underwent the assessment of patients' QoL (McGill QoL Questionnaire), general health status (EQ-5D-5L) and caregiver burden (Zarit Burden Interview [ZBI]). These phone scales were administered in April 2020 (T1) and repeated one month after the end of lockdown (T2), with the addition of a qualitative questionnaire (COVID-QoL-Questionnaire), exploring family reorganization and personal perception of lockdown. Wilcoxon signed-rank test and the chi-squared test were used.

Results

QoL and perceived health status did not worsen during lockdown, while caregiver burden increased ($p = 0.01$). Patient's QoL and caregiver burden were mildly inversely correlated at T1 ($p < 0.05$, rho $= -0.265$); no significant correlations were found at T2. According to the COVID-QoL-Questionnaire, both patients and caregivers were able to consult their physicians while at home (60% and 66.1% respectively). Moreover, caregivers perceived lower family help compared to patients ($p < 0.001$).

Conclusions

In our cohort of ALS patients, QoL and caregiver burden were not compromised by restriction measures during COVID-19 pandemic, while caregiver burden significantly increased. Probably, this is due to the motor impairment in ALS patients, which did not perceive changing in life conditions. Instead, the reported increased burden for primary caregivers could be explained by the restriction of family help, reflecting the importance of a wide social support in ALS patients' management.

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117702

The unfolded protein response in amyotrophic later sclerosis: Results of a phase 2 trial

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Background and aims

Robust preclinical findings demonstrated that guanabenz selectively inhibits ER stress-induced eIF2α-phosphatase allowing misfolded protein clearance, reduces neuronal death and prolongs survival in in vitro and in vivo ALS models.

Methods

In this multicentre RCT with futility design, ALS patients with onset <18 months were randomly assigned to receive in a 1:1:1:1 ratio guanabenz 64 mg, 32 mg, 16 mg or placebo daily for 6 months as add-on-therapy to riluzole. Primary outcome was the proportion of patients progressing to higher stages of disease measured by ALS-MITOs compared to a historical cohort of 200 patients. Secondary