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LETTER TO THE EDITORS

Amyotrophic lateral sclerosis patients' and caregivers' distress and loneliness during COVID-19 lockdown

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Dear Sirs,

The coronavirus (COVID-19) pandemic has influenced psychological and physical health of the general population [1] with chronic patients mostly suffering for public health-care reconfiguration [2]. We aimed at evaluating the impact of this event on a cohort of Italian amyotrophic lateral sclerosis (ALS) patients and caregivers. Patients suffering from respiratory involvement and those with functional disability or rapid progression were expected to have major concern of being infected by COVID-19. To this account, 30 ALS patients and 29 caregivers underwent longitudinal assessment by remote consulting between 18 March and 2 May. None had COVID-19. They compiled questionnaires assessing pandemic distress (Table 1), mood, loneliness, caregiver burden (CBI), and behavioural changes (Table 2). All participants gave informed consent and the local Ethical Committee approved the study. Spearman rho correlations (Bonferroni correction, $p \leq 0.003$) and Mann–Whitney U tests ($p < 0.01$) were performed to verify if COVID-19 distress was related to clinical and neuropsychological profiles. Surprisingly, we did not find significant association between worries of getting COVID-19 and functional disease severity, stage or clinical phenotypes. Instead, it was the feeling of loneliness to play a major role in the fear of getting the infection (rho = 0.672, $p < 0.001$), confirming that social isolation and loneliness are among the most important adverse consequences of the pandemic in ALS patients [3]. Additionally, anxiety was associated with the degree of feelings of being forgotten/rejected by clinicians (rho = 0.543, $p = 0.002$) and change in neurologist–patient relationship (rho = 0.536, $p = 0.003$). Anxious patients were among the most vulnerable ones to suffer from the change of the health-care system. During the pandemic, indeed, many Italian institutions delivered services remotely, possibly coordinating telemedicine activities [4]. However, telematics support for ALS patients should take into consideration the presence of cognitive and behavioural impairment. Between-group comparison revealed that cognitive/behavioural impaired patients, diagnosed according to the consensus criteria [5], did not augment the use of social network to be in touch with relatives and friend during lockdown ($U = 49.5$, $p = 0.009$), possibly due to difficulties in using electronic devices. Moreover, self-rate of behavioural change due to COVID-19 (overall sum of E subscale, Table 1) was inversely related to dimensional apathy scale (DAS) scores (rho = − 0.576, $p = 0.001$), documenting that apathetic patients reported even minor behavioural changes during the pandemic. These findings are in keeping with low awareness in behaviourally impaired patients [5]. Patients' behavioural alterations and motor disability were the greatest burden for caregivers [6], rather than the concern of getting COVID-19 or limitations of daily life due to the quarantine. CBI was indeed associated only with patient disability (ALSFRS-R, rho = − 0.660, $p < 0.001$) and apathy (DAS, rho = 0.557, $p = 0.002$). In caregivers, pandemic distress was documented only as a change in the degree of anxiety possibly due to the greater amount of time spent in the house during lockdown (rho = 0.539, $p = 0.003$). Interestingly, patients and caregivers gave similar answers to the questionnaire on the changes that COVID-19 caused in

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Table 1 Evaluation of the impact of COVID-19 on ALS disease management and daily life

Questionnaire	ALS patients	Caregivers	Between group differences
(A) Perceived risk of COVID-19 (index score)[a]	− 0.254 (1.5)	0.285 (1.8)	n.s
(1) How much are you able to avoid COVID-19?[b]	4.00 (0.8; range 3–5)	3.78 (1.2; range 1–5)	n.s
(2) Amount of information sources on COVID-19 (min 0; max 8)	3.53 (1.3; range 1–6)	.18 (1.5; range 1–6)	n.s
(3) # actions taken to avoid SARS-Cov-2 contagion (min 0–max 9)	8.16 (1.0; range 4–9)	8.44 (0.8; range 6–9)	n.s
(B) Concern about COVID-19			
(1) Worries in the event of an infection [b]	3.53 (1.4; range 1–5)	3.21 (1.6; range 1–5)	n.s
(2) Thinking of COVID-19[b]	3.10 (1.2; range 1–5)	2.96 (1.2; range 1–5)	n.s
(3) Thinking that COVID-19 can worry my family[b]	3.70 (0.9; range 2–5)	3.50 (1.3; range 1–5)	n.s
(C) Change in disease management			
(1) Drug management change[b]	1.30 (0.9; range 1–5)	n.a	–
(2) Change in neurologist-patient relationship[b]	1.96 (1.4; range 1–5)	n.a	–
(3) Feelings of being forgotten/rejected by clinicians[b]	1.46 (0.8; range 1–4)	n.a	–
(4) Concern about negative consequences of COVID-19 healthy emergency on the management of the disease by clinicians[b]	2.65 (1.3; range 1–5)	n.a	–
(D) Change in habits due to COVID-19 state of emergency			
(1) Out-of-home habits[b]	3.56 (1.4; range 1–5)	3.79 (1.2; range 1–5)	n.s
(2) Household habits[b]	1.90 (1.2; range 1–5)	2.21 (1.2; range 1–5)	n.s
(3) Use of social networks[b]	2.23 (1.5; range 1–5)	2.38 (1.3; range 1–5)	n.s
(4) Work/Retirement[b]	2.83 (1.5; range 1–5)	2.62 (1.8; range 1–5)	n.s
(5) Personal care [b]	2.03 (0.9; range 1–5)	1.93 (1.3 range 1–5)	n.s
(E) Change in behaviour due to COVID-19 state of emergency			
(1) Irritable/Nervous[b]	1.66 (1.1; range 1–5)	1.78 (0.8; range 1–4)	n.s
(2) Agitated/Anxious[b]	1.50 (0.7 range 1–4)	1.78 (0.9; range 1–5)	n.s
(3) Sad/Depressed[b]	1.46 (0.7; range 1–3)	1.60 (0.8; range 1–4)	n.s
(4) Bored[b]	1.89 (1.1; range 1–4)	2.03 (1.0; range 1–4)	n.s
(5) Increased consumption of alcohol/cigarettes[b]	1.13 (0.5; range 1–4)	1.25 (0.6; range 1–4)	n.s
(6) Increased food consumption[b]	1.46 (0.8; range 1–4)	1.64 (0.9 range 1–5)	n.s

Data are expressed as means (± standard deviation; range min–max)

n.a. not available data, *n.s.* not significant difference

[a]The perceived risk index was calculated summing z standard scores of the A(1–3) responses

[b]The range of responses varied from 1 (not at all) to 5 (extremely)

daily life. It could be speculated that families accustomed to manage degenerative diseases are more resilient to changes in whatever form they occur. Although the long-term impact of the COVID-19 pandemic is yet to be determined, our study suggests that families with ALS patients, mostly if suffering also form cognitive/behavioural impairment, have higher level of anxiety and feeling of loneliness, and are particularly vulnerable to distress. Tailored interventions should be considered to help them in facing better the changes in habits and health-care system.

Table 2 Demographical, clinical, and psychometric data of ALS patients and caregivers

	ALS patients $N=30$	Caregivers $N=29$	Between group differences
Demographical data			
Age	61.26 (13.0)	56.29 (12.1)	n.s
Male/female	14/16	13/16	n.s
Educational level (min 0; max 9)	3.83 (1.7; range 1–7)	3.53 (1.5; range 1–7)	n.s
Working activities (min 0; max 5)	2.34 (1.0; range 0–4)	2.56 (1.1; range 1–5)	n.s
From Northern/Southern Italy	22/8	22/7	n.s
Clinical data			
Disease duration (months)	24.88 (18.4)	n.a	–
ALSFRS-R	35.93 (4.8)	n.a	–
King's clinical stage (1/2/3 / 4)	7/9/13/1	n.a	–
ALS-MITOS (0/1/2/3/4)	20/10/0/0/0	n.a	–
ALS spinal/ALS bulbar/PLS	13/5/2	n.a	–
ALScn/ALSbi/ALSci/ALScbi	13/4/8/5	n.a	–
Neuropsychological data			
Frontal Behavioural Inventory	5.64 (15.1; range 0–18)	n.a	–
Dimensional apathy scale (DAS)	19.82 (10.6; range 5–47)	n.a	–
HADS-MND anxiety	4.00 (3.25; range 0–10)	n.a	–
HADS-MND depression	2.79 (3.1; range 0–11)	n.a	–
QoL—ALSAQ-5	6.40 (4.1; range 0–16)	n.a	–
QoL—WhoQol-Age	48.23 (10.5)	51.89 (8.0)	n.s
UCLA 3 Items Loneliness Scale	1.10 (1.6, range 0–5)	n.a	–
Caregiver burden inventory	n.a	13.96 (15.1, range 0–60)	–

ALSAQ-5 The Amyotrophic Lateral Sclerosis Assessment Questionnaire (5-item version), *ALSbi* ALS patients fulfilling Strong criteria for behavioural impairment, *ALSci* ALS patients fulfilling Strong criteria for cognitive impairment, *ALScbi* ALS patients fulfilling criteria for ALSci and ALSbi, *ALScn* cognitively-normal ALS patients (i.e.: patients not fulfilling Strong criteria for ALSci and/or ALSbi), *ALSFRS-R* the Revised ALS Functional Rating Scale, *ALS-MITOS* ALS Milano-Torino Staging System, *n.a.* not available data, *n.s.* not significant difference, *PLS* Primary Lateral Sclerosis, *QoL* Quality of Life, *WhoQol-Age* The World Health Organization Quality of Life in the Aging population

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Data availability The datasets generated and analysed during the current study are available from the corresponding author on reasonable request.

Compliance with ethical standards

Conflicts of interest The authors declare that they have no conflict of interest.

Ethical approval The study was performed in accordance with the Declaration of Helsinki and was approved by the ethics board of the IRCCS Foundation "Carlo Besta" Neurological Institute. Written informed consent was obtained from all participants.

Consent for publication All authors declare that the submitted work has not been published before (neither in English nor in any other language) and that the work is not under consideration for publication elsewhere.

References

1. Mazza C, Ricci E, Biondi S, Colasanti M, Ferracuti S, Napoli C, Roma P (2020) A nationwide survey of psychological distress among italian people during the COVID-19 pandemic: immediate psychological responses and associated factors. Int J Environ Res Public Health 17:3165
2. Wang B, Li R, Lu Z, Huang Y (2020) Does comorbidity increase the risk of patients with COVID-19: evidence from meta-analysis. Aging (Albany NY) 12:6049–6057
3. Holmes EA, O'Connor RC, Perry VH, Tracey I, Wessely S, Arseneault L, Ballard C, Christensen H, Cohen Silver R, Everall I, Ford T, John A, Kabir T, King K, Madan I, Michie S, Przybylski AK, Shafran R, Sweeney A, Worthman CM, Yardley L, Cowan K, Cope C, Hotopf M, Bullmore E (2020) Multidisciplinary research priorities for the COVID-19 pandemic: a call for action for mental health science. Lancet Psychiatry 7:547–560

4. De Marchi F, Cantello R, Ambrosini S, Mazzini L, Group CS (2020) Telemedicine and technological devices for amyotrophic lateral sclerosis in the era of COVID-19. Neurol Sci 41:1365–1367
5. Strong MJ, Abrahams S, Goldstein LH, Woolley S, Mclaughlin P, Snowden J, Mioshi E, Roberts-South A, Benatar M, HortobáGyi T, Rosenfeld J, Silani V, Ince PG, Turner MR (2017) Amyotrophic lateral sclerosis—frontotemporal spectrum disorder (ALS-FTSD): Revised diagnostic criteria. Amyotroph Lateral Scler Frontotemporal Degener 18:153–174
6. Burke T, Elamin M, Galvin M, Hardiman O, Pender N (2015) Caregiver burden in amyotrophic lateral sclerosis: a cross-sectional investigation of predictors. J Neurol 262:1526–1532