

Amyotrophic Lateral Sclerosis and COVID-19 – Recommendations to the patients and caregivers

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In patients with Amyotrophic Lateral Sclerosis (ALS) the impact of respiratory infections is expected to be more serious than in the general population, due to the weakness of the respiratory muscles and the limitations in coughing (1-4). ALS patients with impaired respiratory function and/or rapid clinical deterioration are a high risk population for COVID-19, even if no estimates in the different European countries are available.

ENCALS centres and the EAN panel on ALS and Frontotemporal Dementia are working together across countries to provide the best assumed support for our patients to protect them during this pandemic. National guidelines specifically concerning patients with ALS can be lacking and current regulations vary between different European countries. Available National guidelines are as follows, some of them addressing the general population:

Germany: <https://www.dgm.org/aktuelles/aktuelles-meldungen/meldungen/coronavirus-hinweise-covid-19-virus-patientinnen-patienten> (5);

Ireland: <https://hse.drsteevenslibrary.ie/c.php?q=679077&p=4841355>

Italy: <https://aisla.it/covid-19-e-sla-lesperto-risponde/> (6);

Poland: <https://www.gov.pl/web/koronawirus> (7);

<https://gis.gov.pl/aktualnosci/> (8);

Portugal: <https://covid19.min-saude.pt> (9);

UK: <https://www.mndassociation.org/mnd-and-coronavirus/> (10);

<https://www.gov.uk/government/publications/covid-19-guidance-on-social-distancing-and-for-vulnerable-people> (11); <https://t.co/DsgD2EZnXr> (12), with links to <http://medicine.tcd.ie/neurology/> (13); <http://sitran.org/people/shaw/> (14);

Switzerland: <https://www.bag.admin.ch/bag/de/home/krankheiten/ausbrueche-epidemien-pandemien/aktuelle-ausbrueche-epidemien/novel-cov/information-fuer-die-aerzteschaft/schutzmassnahmen.html#72080722> (15);

<https://www.palliative.ch/de/fachbereich/task-forces/fokus-corona/> (16).

In the absence of guidelines specifically written to protect ALS patients and their families/ carers during the COVID-19 pandemic period, and acknowledging the various national regulations the consensus of a European Network for the Cure of ALS (ENCALS) and EAN ALS/FTD group of experts* is as follows:

Recommendations to the ALS patients and family members/ carers, during the pandemic period of COVID-19 (SARS-CoV-2):

1st recommendation: Patients with ALS should be home confined to prevent being infected. If living in a house with a backyard or in the countryside, going out inside the limits of the residency does not present a potential danger. It is

advisable to avoid visitors, including family members/young children, even if symptomless. Hygiene should include more frequent hand washing with soap, avoiding hand-to-face contact (mouth, nose and eyes) and cleaning surfaces with detergent. (17) The carers should sneeze/ cough into the bent elbow or tissue, the tissues should be immediately discarded, and hands should be washed with soap. (17) Mask is required for the patient, family members, or others contacting the patient (18).

2nd recommendation: Identification of the principal carer – The principal carer should coordinate the care provided to the patient. The principal carer should be someone who is always at home with the patient, in accordance with recommendation 1, to prevent being infected outside and transmitting the disease to the patient. Food and medication should be bought through online platforms or by ringing to local shops and having them delivered at home. Mask is required for the principal carer as well (18).

3rd recommendation: Identification of additional carers – If possible, a second carer should be identified. If necessary, the carer (the same per patient) should be available to cover for the principal carer, to prevent having the patient alone during unplanned periods, including due to quarantine/ isolation of the principal carer or others. Mask is required for the additional caregiver (18).

4rd recommendation: Unnecessary care/ Unavailable carer – If self sufficient but living alone without support, or if no carer is available although needed, the patient is advised to contact the caring ALS centre, namely the social assistants who integrate the ALS teams, to report the social situation and to get support if it becomes necessary.

5th recommendation: Outpatient care services – If support is provided by outpatient care services, a principal carer and an additional carer should be identified per patient, limiting external contacts to avoid the risk of spreading the infection. This team, if provided not directly by the ALS team, should keep close contact with the ALS team to report on potential needs/ clinical deterioration/ respiratory emergency. The team should take all necessary measures to prevent a possible transmission of the infection (19).

6th recommendation: In the case of an unplanned period of physical absence from the principal carer and if no additional carer is available, the principal carer or the patient him/herself if capable, should report the situation to the ALS health providers, namely the social assistants integrating the ALS teams, local or national ALS association, palliative care team, homecare institution, local fireman association, or others that support community services.

7th recommendation: The principal carer is responsible for monitoring and providing, in the presence of functional limitations/ dependence and/or fronto-temporal dementia, the basic needs to the ALS patients – feeding/ hydration, administration of medication, respiratory support, mobilization and positioning,

hygiene, comfort, physical and psychological wellbeing. The principal carer should also take care of him/herself, to maintain physical and psychological well-being. It is important to keep timetables, listen to calm and relaxing music, read books, engage in interesting activities at home, have some periods for home physical exercise, if possible. The constant following of the news should be banished.

8th recommendation: If in absolute need to go outside, both patient and carer/ family member should take the advised measures to prevent any risk of infection. Masks may be required, namely FFP2. Check this need with the physician who follows you/ the patient. When arriving home, put off the clothes (coats, bags, scarves, gloves, shoes) in the entrance of the house. Use disinfectant gel containing alcohol. Take off the mask, by only touching the elastics. Wash the hands with disinfectant products containing alcohol (17). If the patient uses walking aids/ wheelchair, these devices should be left also in the entrance of the house. The carer should help the patient. The walking aids/ wheelchair as well as other surfaces, including at the entrance, where the clothes from the outside are gathered, should be disinfected with detergents or products containing alcohol (20). Clothes should be washed with regular laundry detergent with temperatures above 40°.

9th recommendation:

If the patient develops fever, non-productive cough, headache, muscle pain or dyspnoea, in the context of an external contact with someone who could be infected, the patient/ the principal carer should: 1- Stay calm. Do not go to the hospital or other healthcare centres. Stay home; 2- Use masks (both) (17,18); 3- Measure the temperature and write down the symptoms. Have an actualized medication list, including dosing and administration times; 4- Contact the physician or medical team that follows you/ the patient, by phone, mail or available internet platforms.; 5- If no contact is available, phone to the national healthcare line, or the COVID-19 line if available in your country. If nobody answers, ring again. Follow the instructions. If the patient has already been adapted to non-invasive ventilation, a face mask without expiratory holes (or with the expiratory holes closed with tape), connected directly to a filter and to an expiratory piece should be used, to filter the expiratory air. If having a tracheostomy, a filter and the expiratory piece should be connected to the tube. If assistance is needed to remove bronchial secretions, for example by using a mechanical insufflator-exsufflator device, the carer should use FFP2 mask, goggles and gloves, discard all biological material immediately, have a shower immediately afterwards. As the coronavirus can be detected for at least 3 hours after aerosolization, mask and goggles should be used (20).

If the carer/family member is the one developing the symptoms: 1- Stay calm. Do not go to the hospital or other healthcare centres. Stay home; 2-Use masks (both) (17,18); 3- Record your symptoms as well as any symptoms that the patient may have. Have an actualized medication list; 4- Contact the additional carer so that he/she can be prepared to substitute if indicated when phoning the health providers; 5- Contact the COVID-19 line/ the national healthcare line as well as the ALS physician who follows the patient. Follow the instructions.

10th recommendation: In the case of respiratory decompensation, in ALS patients under home ventilatory support or initial respiratory symptoms in the context of ALS, the patient or the caregiver should contact the homecare/ palliative team/ caring ALS centre and inform the physician who regularly cares for the patient. If needed, call the 24/7 health support lines that may have been provided and articulate it with the ALS care centres support. Follow the instructions.

11th recommendation: In case of respiratory emergency, call first the neurologist and pulmonologist in charge to finalize the needed intervention. If needed, call the 24/7 health support lines that may have been provided and articulate it with the ALS care centres support.

12th recommendation: Keep well informed, by reading the news/ listening to reliable sources. Avoid fake news and misleading social media. Rely on the official sources from your country. Limit the time that you spend listening to or reading the news. Be positive. This is a transitory period and it will be solved.

Andrà tutto bene. Vai ficar tudo bem. Wszystko będzie dobrze. Allt ska bli bra. Everything will be alright.

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