

Covid-19 and people with neuromuscular disorders:

World Muscle Society position and advice

The category of neuromuscular disease (NMD) covers a wide range of different diagnoses with widely varying levels of disability even in people with the same diagnosis. It is difficult, therefore to make specific recommendations that apply generally. The following are recommendations that apply to numerous neuromuscular disorders. These recommendations are designed primarily for patients, carers, general neurologists and non-specialist medical providers. They are also intended to inform neuromuscular specialists particularly regarding frequently asked questions and basic service requirements. In-depth reference links are provided.

Note: Covid-19 is a rapidly evolving field. The advice in this document is subject to revision every 3 days. Please ensure that you are using the most up to date version of the document.

1. Are people with neuromuscular disease (NMD) at higher risk?

National neurological associations and neuromuscular networks (Association of British Neurologists, European Reference Network EURO-NMD, others) have produced guidance on the impact of Covid-19 on neurological disorders and their management. These documents define the risk of a severe course of Covid-19 as high or moderately high in all but the mildest forms of NMD. Features conferring a high or very high risk of severe disease include, for example:

- Muscular weakness of the chest or diaphragm, resulting in respiratory volumes less than 60% predicted (FVC<60%), especially in patients with kyphoscoliosis
- Use of ventilation via mask or tracheotomy
- Weak cough and weak airway clearance due to oropharyngeal weakness
- Presence of tracheostoma
- Cardiac involvement (and/or on medication for heart involvement)
- Risk of deterioration with fever, fasting or infection
- Risk of rhabdomyolysis with fever, fasting or infection
- Concomitant diabetes and obesity
- Patients taking steroids and undergoing immunosuppressant treatment

2. What do people with NMD need to do to avoid infection?

Covid-19 spreads through droplet infection when an infected person coughs, sneezes or talks, or potentially via touching a surface carrying infectious droplets. People with NMD and a high risk of a severe course of Covid-19 infection, as defined above, should undertake the following precautions:

- Social distancing of at least 2 metres (6 feet) is a minimum requirement. For high risk individuals (as defined in 1.), self-isolation is advised. Official advice on how to self-isolate should be followed.
- People are encouraged to work from home or stagger their working times if possible.
- Avoid large gatherings and public transport. People in general are urged to limit visits to vulnerable persons.
- Frequent hand-washing (20 seconds with soap and warm water), use of 60% alcohol-based hand sanitizers, and surface disinfection are crucial.

- Caregivers should be in-house, if possible. Essential visiting care givers (for instance, providers of backup support for ventilatory assistance) should wear face masks and adequate PPE according to up to date official guidance, to prevent passing on the virus.
- Visiting physiotherapy is discouraged, however, physiotherapists should provide advice on maintaining physical activity remotely, via phone or videolink.
- It is important to be prepared for all eventualities including when assistants are absent due to illness or quarantine. The person responsible for organizing home care should have an overview of the personnel situation at all times. Plans should be made for how to best meet the needs of the individual without resorting to hospitalization.
- Government advice on protection is regularly updated, and the authors advise patients, carers and medical professionals to follow the up to date recommendations from official websites in their country.

3. What consequences does the risk of Covid-19 infection have for treatments used in people with NMD?

- Patients must ensure they have an adequate supply of medication and of ventilatory support equipment for a period of prolonged isolation (at least 1month supply).
- Patients and carers should make use of online and telephone-based pharmacy and equipment ordering and delivery services.
- Patients and carers need to be comfortable with emergency procedures specific to their condition and their equipment.
- DMD patients on steroid regimens should continue their medication. Steroids must never be stopped suddenly, and there may be a need to increase the steroid dose when unwell.
- Immunosuppression in inflammatory muscle disease, myasthenia gravis, and peripheral nerve disease should not be discontinued except under specific circumstances and in consultation with the neuromuscular specialist.
- Isolation requirements may impact on treatment regimens requiring hospital procedures (i.e. nursinersen (Spinraza), alglucosidase alfa (Myozyme), intravenous immunoglobulin (IVIg) and rituximab infusions or treatments related to clinical trials). These treatments should typically not be stopped, but whenever possible moved to a non-hospital setting (home-visiting or outreach nurses), for which cooperation with manufacturing companies may be negotiated. IVIg can be changed to subcutaneous immunoglobulin whenever possible. Trial centres should be consulted for advice on clinical trials.

4. What needs to be done to assure ventilatory services when isolating (LVR bags, home ventilators etc.)

- Backup and advice hotlines should be offered by the patients' Neuromuscular Centres.
- Patients should have an alert card/medical bracelet providing the Neuromuscular Centre contact.
- Neuromuscular Centres should actively contact patients on ventilatory support to ensure they have relevant information and adequate equipment.

5. When should people with NMD seek admission if they develop symptoms of infection?

Inpatient admission should be avoided if possible, but should not be delayed when necessary. This can be a difficult decision. People with NMD need to be aware that:

- Emergency services may be under severe pressure.
- Individual countries may have triaging procedures in place. These may affect the potential for intensive care admission for people with NMD who require ventilation. Specifically, the terms “incurable” and “untreatable” may be confused by medical staff. Neuromuscular disorders may be incurable, but they are not untreatable, and the implications for treatment decisions are very different.
- Use of patients’ home equipment (i.e. ventilators) may be prohibited by some hospital infection-control policies, or require modifications. Ideally, there should be a back-up plan.

6. Can treatments for Covid-19 have effects on neuromuscular disease?

- Numerous specific treatments for Covid-19 are under investigation. Some of these can affect neuromuscular function significantly: for example, chloroquine and azithromycin are unsafe in myasthenia gravis, except when ventilatory support is available.
- Other treatments may have effects on specific neuromuscular diseases (in particular, metabolic, mitochondrial, myotonic and neuromuscular junction disorders), and anatomical peculiarities may influence options for treatment (e.g. prolonged prone ventilation)
- Experimental treatments for Covid-19 may be offered “compassionately”, i.e. outside trial conditions. They should only be taken after consultation with the patient’s neuromuscular specialist.

7. What should neuromuscular specialists do to assist Emergency Medical and Intensive Care decisions on admission to units, escalation of treatment, and ceilings of care in neuromuscular patients?

Decisions on patient admission to Intensive Care may be affected by anticipated or existing capacity problems. Triaging may have been instituted. This can have practical and ethical consequences.

- There must be close collaboration between neuromuscular and respiratory physicians.
- The neuromuscular specialist must be available to play a role in ensuring fair provision of intensive care to NMD patients.
- Ideally, neuromuscular specialists will have involved themselves in formulating hospital policies, decision-making algorithms and documentation forms.
- Neuromuscular specialists must develop guidelines for treatment that ensure patients remain at home as long as possible.

8. What patient support should neuromuscular centres provide?

Neuromuscular centres and specialist services should aim to provide the following:

- Patient hotlines staffed by neuromuscular care advisors, physiotherapists and other specialist personnel, with specialist physician backup (paediatric and adult).
- The possibility to continue routine clinics by structured telemedical phone and video links (for this, national and institutional data security regulations such as HIPPA approval may need modification).
- Outreach ventilatory support strategies should be provided.
- Strategies to maintain hospital-based treatments with minimal disruption.

- Neuromuscular specialists should be in discussion with their hospitals' Emergency, Medical and Intensive Care departments on restrictions for use of home NIV equipment.
- Neuromuscular specialists should support their hospital to define approved devices and ensure their availability (i.e. ICU mask systems with viral particle filters to permit use of patients' NIV machines in hospital).
- Liaison and shared care with Intensive Care services.

Further information:

Documents are available at

<https://www.theabn.org/page/COVID-19>

<https://neuromuscularnetwork.ca/news/covid-19-and-neuromuscular-patients-la-covid-19-et-les-patients-neuromusculaires/>

<https://www.youtube.com/watch?v=3DKEeRV8alA&feature=youtu.be>

<http://www.eamda.eu/2020/03/19/coronavirus-covid-19-information-for-people-with-nmd/>

<https://www.gov.uk/government/publications/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19>

<https://ern-euro-nmd.eu/>

European Neuromuscular Centre website: www.enmc.org

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