

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 who have been diagnosed with a neuromuscular disorder, their 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 regular revision. Please ensure that you are using the most up to date version of the document.

Update (28-11-2020): The WMS have considered the advice provided in this document in light of the new wave of infections. The recommendations made in previous versions still hold true.

Amendments suggested through newer experience are provided at the end of each paragraph and are clearly marked.

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

So far, there is no evidence that hereditary neuromuscular disorders confer a higher risk of infection by the SARS-CoV-2 virus. However, neuromuscular diseases and their treatments may influence the patient's ability to cope with infection or its systemic effects.

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)
- Conditions with a risk of deterioration with fever, fasting or infection (eg. neuromuscular junction or metabolic disorders)
- Conditions with a risk of rhabdomyolysis with fever, fasting or infection
- Frailty, concomitant diabetes and obesity and severe hypertension
- Patients taking steroids and undergoing immunosuppressant treatment

The following update (28-11-2020) applies:

- *The risk associated with steroid treatment may be less than initially feared. Patients taking steroids and undergoing immunosuppressant treatment should continue their treatment and need to be carefully monitored throughout the pandemic.*
- *Frailty and hypertension are recognised as risks.*

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 1.5-2 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. Decreasing infection risk may allow gradual de-escalation.
- 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 (FFP2/N95 or FFP3) and adequate PPE according to up to date official guidance, to prevent passing on the virus.
- Physiotherapists should provide advice on maintaining physical activity remotely, via phone or videolink where suitable. In physiotherapy sessions that require a home visit, an adapted protective setting has to be ensured (FFP2/N95 or FFP3 mask for the physical therapist and the carer present in the room, surgical or FFP2 mask for the patient, protective clothes, gloves and glasses or helmet should be used by the physical therapist).
- 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.

The following updates (28-11-2020) apply:

- *Physiotherapy is important maintenance for neuromuscular patients and the updated advice has moved away from blanket recommendation against home visits, where safety procedures can be strictly observed.*
- *The WMS re-emphasises its advice to work from home where possible, and people with neuromuscular disorders at elevated risk should not resume office or customer contact work without seeking medical advice from their Neuromuscular Centre.*
- *People with neuromuscular disorders are advised to wear face masks (surgical masks) when leaving the home; higher grade masks may be considered in high risk areas.*

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 1 month 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 pre-emptively except under specific circumstances and in consultation with the neuromuscular specialist.
- Whether and when to start new immunosuppressive treatment may be influenced by how severe the risk of infection is perceived, versus risks of deferring treatment.
- 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 moving treatment to a non-hospital setting should be considered (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.

The following update (28-11-2020) applies:

- *Where treatments cannot be safely continued, or where services are disrupted, the Neuromuscular Centre can provide advice on safe duration of delays to treatment, and on effective “catch up” schemes.*

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.

The following update (28-11-2020) applies:

- *On attending hospital, people with neuromuscular disorders should bring with them detailed advice on use of their medical devices (eg. home ventilator) in the hospital setting provided by manufacturers and neuromuscular networks.*

6. What applies to immunosuppressive treatment in patients who have suspected or proven COVID-19 infection?

- The decision to temporarily withhold immunosuppressant medication, or change to a different agent must be made in the individual situation, with the neuromuscular specialist.
- Steroid treatment should not be withheld or stopped.
- IVIg, plasma exchanges, and complement inhibitor treatment such as Eculizumab are not expected to affect the risk of COVID-19 infection or of severe disease.

7. 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. Cardiotoxicity and QT-prolongation through chloroquine and hydroxychloroquine can potentially worsen cardiomyopathy.
- 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.
- So far, we are not aware of trials of live virus vaccines, where there could be a risk for immunosuppressed patients (<https://www.who.int/publications/m/item/draft-landscape-of-covid-19-candidate-vaccines>). However, this is an area in rapid development and WMS intend to provide separate information in the near future.

The following updates (28-11-2020) apply:

- *The use of Hydroxychloroquine is actively discouraged following evidence for no benefit*
- *Evidence-based treatment for COVID-19 such as dexamethasone, hydrocortisone and/or remdesivir can be applied in people with neuromuscular disorders, if needed through the severe phase of COVID-19*
- *Vaccines: This is a rapidly evolving topic. A number of vaccines are approaching approval, and WMS intends to provide separate information in the near future as vaccines gain approval for clinical use. The WMS note that people with neuromuscular disorders constitute a medical risk group qualifying for early vaccination, in most official documents world-wide.*

8. 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. Triage 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. Patients should not be labelled as “terminal” and triaged for non-treatment simply on the basis of their disability and diagnosis.

- 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.

9. 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).
- Support through routine specialist clinics should be continued through remote monitoring using structured telemedical phone and video links. Multiple, nationally approved platforms are now available with guidance from specialist societies.
- Many clinical assessments such as swallowing tests can be done remotely by video link.
- 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.
- Provide advice on rehabilitation in the home for neuromuscular patients, including Telehealth approaches
- Facilitation of mask and PPE supply for patients and carers

10. De-escalation of shielding measures – “Déconfinement”

Since the last update of these recommendations, a new discussion is emerging, concerning the safe de-escalation of some aspects of shielding and self-isolation to allow people with neuromuscular disorders to resume social interactions and education, work, or attend scheduled medical appointments.

Currently, there is considerable variation in the way national restrictions and the relaxation of restrictions are evolving, and this impacts on people with neuromuscular conditions, their families and carers, who seek advice from their neuromuscular services.

Despite the range of national approaches to this issue, the WMS agrees on the following considerations regarding risk stratification:

- For people with neuromuscular disorders who are considered at low risk (see paragraph 1 of the Advice and Position document), i.e. no cardiorespiratory impairment, no immunosuppression, and no significant risk-elevating factors and comorbidities, we suggest the cautious following of local and national guidance. In doubt, consultation with the neuromuscular specialist is recommended.
- For patients with medium risk, for instance with mild respiratory involvement, we advise a detailed discussion with their neuromuscular specialist, or with the physician responsible for their neuromuscular care. Controlled relaxation of restrictions, strictly in a secure environment, may be considered, with appropriate caution and taking into account local and national recommendations.

- Special considerations must remain for people considered at “high” or “very high” risk (see paragraph 1 of the original document), in particular people with severe or unstable respiratory compromise (FVC < 60% predicted); reliance on home ventilation; clinically relevant impairment of heart function; immunosuppression; or severe weakness requiring multiple carers or complex ongoing support. For these patients, measures to avoid infection including self-isolation should remain in place. Carers and family members who are no longer in self-isolation must continue to use masks and barriers when in contact with the person at risk.
- For children and adolescents with neuromuscular disorders, and their parents, the major question will be whether a return to school and child care centres is acceptable. The safety of children with neuromuscular disorders will also be a concern for the schools and childcare centres these children attend. The decision to return will depend on individual factors, regarding both the individual and institution, but details such as staffing levels and hygiene protection according to national guidelines, will be important. Some recommendations are already available (see References); internationally there is significant variation.
- Patients also need to be reassured that they can safely attend hospitals for important procedures such as sleep studies, cardiac tests, and initiation of non-invasive ventilation. Neuromuscular specialists need to monitor their patient cohort to detect what medical procedures or monitoring may have been postponed during “lockdown”, and begin arrangements for these services to be safely resumed. Neuromuscular services should ensure that their hospital is making adequate provisions for the safety of people with neuromuscular disorders attending, including staff adequately equipped with personal protection, designated “green” areas in hospitals for non-COVID-19 related treatments, and safe waiting areas and consulting rooms with appropriate distancing between patients and staff.

The following update (28-11-2020) applies:

- *Currently, the majority of Northern Hemisphere countries are re-introducing more restrictive measures, and in part, “lockdown”. People with neuromuscular disorders should only follow de-escalation measures where up to date and approved by their Neuromuscular Centre.*

Experience so far (15-06-2020): Based on an unpublished survey among WMS members, we urge particular caution in some patient groups, despite their being ambulatory and socially active. These may include patients with conditions that are likely to exacerbate through febrile illness, and those with prominent nasopharyngeal weakness. In our case observations worldwide, Myasthenia gravis and Myotonic dystrophy have featured prominently in this group.

The following updates (28-11-2020) apply:

- *This experience is broadly confirmed by newer data*
- *Potential long-term consequences of COVID-19 among people with neuromuscular disorders are as yet unclear*

Further information links:

Note (28-11-2020): unapproved Internet and social media sites may contain inaccuracies and disinformation. Ensure that inline information is directly sourced from official sites

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

<https://neuromuscularnetwork.ca/news/COVID-19-and-neuromuscular-patients-la-COVID-19-et-les-patients-neuromusculaires/> (updated version April 3rd 2020)

<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/>

<https://www.enmc.org> (European Neuromuscular Centre website)

<https://www.aanem.org/Practice/COVID-19-guidance> (American Association of Neuromuscular and Electrodiagnostic Medicine AANEM website)

<https://www.apta.org/telehealth> (American Physical Therapy Association advice on telehealth)

<https://filnemus.fr> (French neuromuscular reference centres network with extensive advice on medical and social issues around Covid19)

http://www.filnemus.fr/menu-filiere/evenements/actualites/article/news/les-recommandations-de-filnemus-dans-la-periode-du-deconfinement/?tx_news_pi1%5Bcontroller%5D=News&tx_news_pi1%5Baction%5D=detail&cHash=56f89313ee508b50526c7dbfb952dd7a

<https://www.nvk.nl/over-nvk/vereniging/dossiers-en-standpunten/covid-19/document-covid-19?dossierid=26542080>

Reference update 28-11-2020, Vaccines against COVID-19, and neuromuscular disorders:

<https://www.who.int/publications/m/item/draft-landscape-of-covid-19-candidate-vaccines>

<https://www.mda.org/press-releases/facebook-live-qa-covid-19-vaccine-development-and-impact-clinical-trials>

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