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Practice guidelines

Guidance for the care of neuromuscular patients during the COVID-19 pandemic outbreak from the French Rare Health Care for Neuromuscular Diseases Network

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ABSTRACT

In France, the epidemic phase of COVID-19 caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) began in February 2020 and resulted in the implementation of emergency measures and a degradation in the organization of neuromuscular reference centers. In this special context, the French Rare Health Care for Neuromuscular Diseases Network (FILNEMUS) has established guidance in an attempt to homogenize the management of neuromuscular (NM) patients within the French territory. Hospitalization should be reserved for emergencies, the conduct of treatments that cannot be postponed, check-ups for which the diagnostic delay may result in a loss of survival chance, and cardiorespiratory assessments for which the delay could be detrimental to the patient. A national strategy was adopted during a period of 1 to 2 months concerning treatments usually administered in hospitalization. NM patients treated with steroid/immunosuppressants for a dysimmune pathology should continue all of their treatments in the absence of any manifestations suggestive of COVID-19. A frequently asked questions (FAQ) sheet has been compiled and updated on the FILNEMUS website. Various support systems for self-rehabilitation and guided exercises have been also provided on the website. In the context of NM diseases, particular attention must be paid to two experimental COVID-19 treatments, hydroxycholoroquine and azithromycin: risk of exacerbation of myasthenia gravis and QT prolongation in patients with pre-existing cardiac involvement. The unfavorable emergency context related to COVID-19 may specially affect the potential for intensive care admission (ICU) for people with NMD. In order to preserve the fairest medical decision, a multidisciplinary working group has listed the neuromuscular diseases with a good prognosis, usually eligible for resuscitation admission in ICU and, for other NM conditions, the positive criteria suggesting a good prognosis. Adaptation of the use of noninvasive ventilation (NIV) make it possible to limit nebulization and continue using NIV in ventilator-dependent patients.

1. Introduction

Medical practices worldwide have faced unique challenges in the context of the ongoing COVID-19 outbreak. Among the various existing medical disciplines, the pandemic has significantly changed the current practice in treating neuromuscular disorders. These diseases constitute a group of very heterogeneous conditions, most often of genetic or autoimmune origin, which affect both children and adults to a degree that varies widely from one individual to another. They include muscle disorders (e.g., muscular dystrophies, congenital myopathies, metabolic myopathies, inflammatory myopathies, and muscle channelopathies), diseases of the neuromuscular junction (e.g., either acquired or congenital myasthenic syndromes), peripheral nerve disorders (e.g., dysimmune neuropathies, familial amyloid neuropathies, and Charcot–Marie–Tooth disease), and spinal muscular atrophies. In France, 40,000–50,000 patients are thought to suffer from neuromuscular diseases. A significant number of these patients display great disability and may have cardiac and/or respiratory impairments. Of concern for this report, this number excludes those with amyotrophic lateral sclerosis; although also included under the umbrella of neuromuscular disorders, the herein proposed guidance will not cover this condition.

In France, the epidemic phase of COVID-19 caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) began in February 2020 and resulted in the implementation of emergency measures and a degradation in the organization of neuromuscular reference centers. The regional health agency for Île-de-France provided recommendations for COVID-19 support in neurology [1]; the main challenge was to protect patients with neurological pathologies from contamination and to organize the continuation of necessary treatments in view of maintaining the continuity of care. The French Rare Health Care for Neuromuscular Diseases Network (FILNEMUS) also announced guidance in an attempt to homogenize the management of neuromuscular patients in this context to limit the contamination of extremely fragile patients while avoiding the loss of survival chance linked to the interruption of essential treatment or follow-up.

In the uncertainty expanding with the evolution of the epidemic, national measures have been proposed by FILNEMUS to last for a period of 2 months (as of the time of writing of this manuscript, from March to April 2020) to homogenize health care in France. However, it is difficult to establish specific guidelines concerning the heterogeneity of diseases and of patients being managed for the same disease. Different scenarios considering regional specificities, particularly in terms of the severity of the epidemic, are then considered and will be the subject of subsequent guidance with the view of offering optimal care to our patients in accordance with our public health responsibilities.

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2. General recommendations relating to COVID-19 support in neurology

For neurological pathologies being assessed or requiring treatment in health care establishments, in cases where there is no emergency or risk to suspending treatment, one recommendation is to deprogram hospitals for a period of time. Furthermore, to prepare to take care of patients suspected or confirmed to have COVID-19, suspected or confirmed COVID-19 patients must be separated from non-COVID-19 patients by ensuring reinforced hygiene rules and by applying barrier measures (among both patients and staff).

Conducting telephone interviews when the patient makes an appointment is important in eliminating signs of COVID-19 (e.g., fever, chills, high or low respiratory signs, and body aches). If possible, surgical masks should be worn by all patients with comorbidities upon their arrival at the hospital. Patients presenting for treatment of neuromuscular conditions should be placed in a non-COVID-19 hospital sector/service, taking all necessary measures to avoid contamination.

If a patient with a neuromuscular condition also shows symptoms of COVID-19 or their test results are positive, advice from the neurologist concerning the unique measures to be applied should be considered. In the former case, the diagnosis of COVID-19 should be confirmed by conducting polymerase chain reaction testing. If necessary, the patient should be referred to a COVID-19-specific health unit to protect the nursing staff at the hospital.

3. Delivering information to and communicating with patients

Patients with neuromuscular diseases may have risk factors for developing severe forms of COVID-19 and receiving prolonged intensive care may worsen their functional prognosis. It is therefore essential to develop a preventive approach. This involves the supply of easily intelligible information to this patient population, the content of which must be updated as the knowledge progresses and widely disseminated. As patients demand information to keep themselves informed and safe and to homogenize the answers at the national level, a frequently asked questions (FAQ) sheet has been compiled from the questions most frequently asked of the secretariats at reference centers; the coordinator nurses; and the regional offices of the AFM-Telethon, French neuromuscular patient association. The main addressed topics pertain to the disease itself, its mode of transmission, the official national public health recommendations to prevent it, the behaviors to adopt in case symptoms appear, and its consequences specific to the neuromuscular patient (e.g., relating to the management of steroids and immunosuppressor treatments, medical appointments, rehabilitation, and ventilation). The constant evolution of both available knowledge and official instructions have made it necessary to continuously update the FAQ content. Achieving the widest possible dissemination is necessary for this type of action to be effective. This FAQ sheet is therefore available on the FILNEMUS website (http://www.filnemus.fr/), and its dissemination is ensured by the neuromuscular reference centers and the AFM-Telethon in particular, thanks to the offering of backup and advice hotlines by the reference centers or the regional offices of the AFM-Telethon. In addition to fulfilling the mission of disseminating information, these hotlines offer a way to connect with patients and identify risky situations such as those involving the loss of human aid, isolation, disruption of care, or psychological distress. The reference center should therefore ensure that patients have received relevant information and adequate equipment. Within the reference centers, these hotlines can be managed by paramedical staff working from home but also by residents redeployed from other medical units whose normal activities have been reduced.

4. Appointment management and regular follow-up

To reallocate human and material resources in the fight against COVID-19, nonurgent appointments should be postponed or, if appropriate, replaced by telemedicine [2,3]. Teleconsultation is only possible by videoconferencing, and administrative steps have traditionally slowed down its development (decree No. 2010-1229 of October 19, 2010, on telemedicine), but the rules have since been relaxed: when the patient does not have the necessary equipment to perform videoconferencing, telecare activities can be conducted by phone (decree No. 2020-227 on March 9, 2020). A register of canceled appointments must be kept by the reference center to reschedule at a later date. Acts such as electroneuromyography and muscle and nerve biopsies must be reserved for diagnostic emergencies such as in the case of vasculitis, Guillain–Barré syndrome, myasthenia gravis, or myositis.

Hospitalization should be reserved for emergencies, the conduct of treatments that cannot be postponed (poorly balanced condition, relapse), check-ups for which the diagnostic delay may result in a loss of survival chance, and cardiorespiratory assessments for which the delay could be detrimental to the patient. Similar to with canceled appointments, a register of canceled hospitalizations must be kept by the centers for rescheduling to a later date. Teleconsultations conducted by a doctor, a resident, or even a coordinator nurse will easily be able to verify that the cancelation of the hospitalization will not harm the patient.

5. Management of the interruption of physiotherapy support

Maintaining joint flexibility, muscle strength, and endurance is recommended in many neuromuscular diseases through regular in-house or office care by health professionals [4–8]. In March 2020, a decree regulating mobility to increase the resistance against the spread of the COVID-19 led to the closure of private therapists' practices, including those of physiotherapists, speech therapists, and occupational therapists (decree No. 2020-260 on March 16, 2020). A subsequent recommendation of the council of the order of physiotherapist

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called for ''the maintenance of in-house physiotherapy of vulnerable patients for whom the cessation of care could lead to a major aggravation, noting that the usual management of many patients is severely disrupted. During the period of outbreak, the High Council of Public Health recommends for the development of activities allowing the continuity of patient support when it is feasible at a distance (for example, by tele-education) [9].

A working group of FILNEMUS has since set up various support systems (e.g., illustrated sheets with links to online videos) for self-rehabilitation and exercises performed by caregivers or relatives. They are available on the FILNEMUS website (http://www.filnemus.fr/) and free of copyright and have been validated previously. Classified by theme (e.g., according to pediatric or adult age, degree of motor impairment, and objectives), these exercises are deliberately simple and guided so that they can be performed by people who are not health professionals. It is suggested that these exercises be adapted to individual situations under the control of the referring doctor and in close collaboration with the usual therapists. As most of the neuromuscular reference centers and competence centers have maintained significant teleconsultation activity, patients and families are encouraged to contact these departments to receive individualized support.

6. What to do about treatments usually administered during hospitalization

In the epidemic context, just prior to the containment period, a national strategy was adopted concerning treatments usually administered in hospitalization. This strategy was developed only for implementation during a period of 1 to 2 months and will need to be adopted further thereafter according to the evolution of the epidemic in the region.

6.1. *Treatment with recombinant human GAA (rhGAA) (Myozyme®) for Pompe disease*

Currently, 115 adult patients have been treated with alglucosidase alfa (Myozyme®; Genzyme Corporation, Cambridge, MA, USA), according to the French registry for Pompe disease, 20 of whom were included in the neoGAA and COMET (Sanofi, Paris, France) and PROPEL (Amicus Therapeutics, Cranbury, NJ, USA) clinical trials.

Treatment with these enzyme replacement therapies, often administered during daily hospitalization, has been discontinued in most cases since the beginning of the epidemic to limit the risk of patients with Pompe disease contracting COVID-19 while in the hospital, especially since most patients with Pompe disease have respiratory insufficiency related to diaphragmatic involvement. Stopping enzyme replacement therapy over a period of 1 to 3 months is unlikely to lead to a significant worsening of the disease, although there are few data available in the medical literature with which to accurately estimate the risk of worsening the disease after a relatively short interruption of treatment. A Swiss study of seven patients with Pompe disease whose treatment was interrupted for economic reasons [10] showed clear deteriorations of motor and respiratory functions and no complete recovery following the resumption of treatment [11]. However, in this study, the duration of treatment interruption was greater than 9 months in six of seven cases, and there was no clinically significant worsening in the remaining patient having the shortest duration of interruption of 3 months.

Assuming that many of the daily in-patient units will remain closed for several weeks or months, the possibility of home infusions could be considered to limit the risk of contamination of patients in the hospital in the short term and to relieve the day hospital structures that will have to deal with an influx of patients during the course of the epidemic. When treatment is resumed, the evaluation of motor and respiratory functions should be performed, with pulmonary function and 6 min walking test conducted to assess the impact of this suspension of infusions.

6.2. *Intrathecal injections of nusinersen for spinal muscular atrophy*

The initiation of nusinersen represents a therapeutic emergency for children with type 1 or type 2 spinal muscular atrophy (SMA). The indication should be maintained because of the functional and vital consequences of delaying treatment. For adolescents and adults with type 2 SMA and type 3 SMA, the therapeutic objective is to stabilize or slightly improve the functional state. Therefore, for these patients, the initiation of treatment may be delayed. For patients already under treatment, nusinersen has a tissue half-life of more than 100 days with an effect on alternative splicing present for several months [12]. Moreover, there appears to be a cumulative phenomenon as intrathecal concentrations of nusinersen dosed prior to injection increased on average from 1.68 ng/mL at 15 days after the first injection to 7.46 ng/mL at 4 months after the sixth injection [13]. It is recommended to try as much as possible to continue intrathecal injections in patients with type 1 SMA and among young children with type 2 SMA. For adolescents and adults, injections could be delayed by 1 to 4 months depending on the evolution of the disease. At this time, for patients with arthrodesis, injections guided by computed tomography are no longer available in most university hospitals because of the reorganization of services to combat the COVID-19 pandemic.

After the COVID-19 outbreak is brought under control, several dose catch-up schemes are possible as follows:

- rapid catch-up for the most progressive forms:
  - if the dose is delayed by less than 3 months, the treating physician should inject as soon as possible and restart the schedule at the eighth month initially planned,
  - if the dose is delayed by 4 months, the treating physician should inject two doses 15 days apart;
- slow catch-up for the less progressive forms:
  - if the dose is delayed by less than 3 months, the treating physician should inject as soon as possible and continue every 4 months thereafter until the end of treatment,
  - if the dose is delayed by 4 months, the treating physician should entirely restart the treatment regimen and dose every 4 months.

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6.3. *Treatment by patisiran for familial amyloid neuropathy (FAP)*

It is recommended to interrupt the delivery of patisiran infusions in the hospital to avoid exposing these patients to the risk of COVID-19 contamination in the hospital environment as these individuals are considered to be at high risk for serious complications (e.g., due to age of older than 70 years and underlying cardiomyopathies) with a relay conducted by infusions performed at home. The time necessary to organize the relay and the interruption of one to two doses should not pose any particular long-term problem.

A phase II study showed that the injection of patisiran at 0.3 mg/kg reduced serum TTR very rapidly by more than 80% over a 3-week period [14]. In phases II and III extension clinical trials (APOLLO), the interruption of one to two infusions of patisiran, authorized in the protocol, among several patients, did not have a negative impact on the final results [15]. Moreover, no anti-small interfering RNA antibodies were observed in any of the treated patients.

6.4. *Intravenous immunoglobulin*

The treatment of dysimmune neuropathies (e.g., chronic polyradiculoneuropathy, Lewis–Sumner disease, and multifocal motor neuropathy with conduction blocks) most often involves the iterative administrations of either intravenous or subcutaneous immunoglobulins, performed in the hospital or at home, the benefit of which has been widely demonstrated [16]. Interruption of this therapy may result in the re-exacerbation of the symptoms of neuropathy; thus, home therapy should be organized. However, unfortunately, this arrangement is not always possible, given enhanced staff shortages, and is unsuitable for treatment initiation. Switching to oral corticosteroids [16] is not recommended during SARS-CoV-2 infection and may worsen the disease in dysimmune motor neuropathies [17]. In myasthenia gravis, intravenous immunoglobulin is also used, either during an outbreak or recurrently [18]. In the latter case, the recommendation for home therapy was also made, and the same is true in the context of dysimmune neuropathies unless the severity of the relapse warrants in-patient monitoring.

7. Guidelines for immunosuppressive and immunomodulating treatments for dysimmune pathologies

The FILNEMUS network recommends that patients treated with immunosuppressants for a dysimmune pathology should continue all of their treatments in the absence of any manifestations suggestive of COVID-19. It should be reminded that stopping treatment may lead to a relapse of the disease. Particular attention should be paid to steroid therapy, since the abrupt interruption of corticosteroid therapy may be responsible for acute adrenal insufficiency. As the data in the literature are constantly evolving, these guidelines will be periodically re-evaluated.

There is no contraindication in the context of COVID-19 to initiating immunosuppressant treatment (e.g., azathioprine, methotrexate and mycophenolate mofetil) to control a severe inflammatory pathology. This would, of course, be accompanied by a strict application of public health recommendations. For methotrexate more specifically, the switch from the injectable to the oral route can be discussed to prevent the patient from being contaminated by the home care nurse. Ultimately, the decision depends on the reason for choosing the injectable form. If it is a reason of efficacy, then optimal disease control is necessary. Conversely, if there is a problem of tolerance, it may be difficult to reverse the decision, and a dose-splitting trial may be warranted in this regard.

As for biotherapies (e.g., rituximab and equivalents), it is justified to maintain them if they are effective and well-tolerated to avoid the potential occurrence of a relapse, in conjunction with strict application of public health recommendations. For the initiation of a biomedication, decisions will have to be made on a case-by-case basis depending on the pathology (e.g., anti-MuSK myasthenia gravis), its severity, the regional situation of the epidemic, and the informed consent of patients [19].

In the case of COVID-19 infection, steroids should not be stopped abruptly because of the risk of adrenal insufficiency. High-dose steroid therapy has been tested previously in respiratory syndromes such as SARS or MERS without worsening or even improving mortality rates [20,21]. Currently, data about the effects of other treatments remain very sparse; nevertheless, the first results from the Italian experience are demonstrative. Out of a cohort of 320 patients with chronic inflammatory rheumatic diseases treated by biotherapies or JAK inhibitors, eight patients had a COVID-19 infection either confirmed or strongly suspected [22]. In all cases, the authors reported that they suspended the treatments during the infection and resumed them after a ''transient'' discontinuation. Any discontinuation of immunosuppressive therapy should be discussed with the referral center following the patient. As in the experience of infectious events, particularly bacterial events under biotherapy, the resumption of immunosuppressant therapy should be discussed 1 to 2 weeks after the absence of any symptoms, i.e., a total cessation of around 3 to 4 weeks from the start of COVID-19 infection. Azathioprine cannot be crushed and then cannot be administered if the patient is tube-fed. In this case and if the decision to maintain immunosuppression is made, a switch to mycophenolate mofetil syrup can be an option to consider. Finally, biotherapies such as tocilizumab are promising candidates for the treatment of the cytokine storm responsible for COVID-19 acute respiratory distress syndrome [23].

8. What to do about cardiac treatments

Some patients have been tempted to stop their angiotensin converting enzyme (ACE) inhibitors or angiotensin II receptor blockers (ARBs), given the release of information (mostly transmitted on social media), suggesting that these drugs may increase both the risk and severity of COVID-19 infection. This concern arose from the observation that the virus binds to ACE2 to infect cells and the fact that ACE2 levels are increased in patients treated with ACE inhibitors and ARBs. However, there are currently no data proving a causal relationship

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between ACE2 activity and COVID-19–associated mortality [24]. Some findings from preclinical studies even suggest a possible protective role of ARB in COVID-19-associated lung injury [25]. It has been well documented that the abrupt withdrawal of ACE inhibitors or ARBs in high-risk patients, including those with cardiac dysfunction, may result in further clinical instability and adverse health outcomes. Thus, patients who are at risk for, being evaluated for, or who have been diagnosed with COVID-19 should continue with their usual cardiac treatments, including ACE inhibitors and ARBs [26].

9. Conduct of therapeutic trials

In the epidemic context, it was decided over a period of 1 to 2 months to discontinue inclusions in clinical research protocols. Home delivery of products to those already enrolled was arranged with study sponsors, and follow-up visits were converted into teleconsultations. However, visits and treatments that are essential for urgent clinical care may concurrently still serve as research visits, in the appropriate circumstances.

10. Particularities of neuromuscular patients concerning treatments prescribed for COVID-19

Numerous therapeutic trials have been initiated in the context of COVID-19 infections. As one example, a large randomized European study (DISCOVERY) evaluating four treatments for COVID-19—specifically, remdesivir, lopinavir in combination with ritonavir, ritonavir in combination or not with beta interferon, and hydroxychloroquine—was started in March 2020 (ClinicalTrials.gov identifier No. NCT04315948). Although various treatments under review appear promising, their effectiveness still needs to be confirmed in larger populations. At this time, certain experimental treatments for COVID-19 may be offered ''compassionately,'' i.e., outside trial conditions. In the case of patients with neuromuscular disorders, the medication should only be taken after consultation with the patient's neuromuscular specialist.

In the context of other neuromuscular diseases, particular attention must be paid to two experimental COVID-19 treatments, hydroxycholoroquine and azithromycin [27]. A retrospective study of 127 patients with autoimmune myasthenia gravis showed a worsening of the disease after taking treatments such as azithromycin (odds ratio: 1.42), fluoroquinolone (odds ratio: 0.89), and beta-blockers (odds ratio: 2.70). These treatments are also contraindicated in congenital myasthenia gravis [28]. However, if the patient is intubated and ventilated, these antibiotics can be used in the absence of alternative therapy. It should be kept in mind that the worsening or flare-up of myasthenia gravis usually occurs within days of the administration of the contraindicated antibiotic treatment, but flare-ups may occur later on up to 29 days after the initiation of treatment [29]. In the absence of a demonstrated benefit from hydroxychloroquine and given that an exacerbation of myasthenia gravis may be caused by this molecule, although reports are contradictory [30,31], the FILNEMUS network does not recommend their use in myasthenic patients with COVID-19 infection at the time of writing.

For other neuromuscular diseases (and, in particular, Andersen's syndrome), great attention must be given to the fact that the primary cardiac effect of hydroxychloroquine is QT prolongation, which may lead to cardiac arrest secondary to cardiac arrhythmia. This risk is increased when hydroxychloroquine is combined with azithromycin, which has the same cardiac effects. The decision to initiate or continue with this treatment can only be made based on an individual benefit–risk assessment, considering:

- corrected QT measurements and the occurrence of cardiac arrhythmia;
- the effectiveness of the treatment on COVID-19 infection;
- the presence of QT-modulating factors (e.g., ionic disturbances such as hypokalemia or QT-prolonging drugs).

There is also an increased risk of conductive disorders and systolic left ventricular dysfunction, which have been mainly reported for long-term treatments.

11. Recommendations for emergency management of neuromuscular patients in the COVID-19 setting

The COVID-19 pandemic, unprecedented in terms of the number of patients affected and its severity, has prompted growing concerns about the capacity of the various health systems to cope with the resultant increased medical needs, particularly in terms of having adequate resuscitation beds. In France, the excess of patients in the intensive care unit (ICU) has led to transfers of patients from the Greater East region and Paris to other regions in France or even other neighboring European countries. In France, the medical community—including, particularly, ICU practitioners—has raised the possibility of having to make difficult choices and prioritize patients concerning access to resuscitation in an emergency. Age and the presence of comorbidities appear to be the risk factors for developing severe COVID-19 infection. Recent papers also reported that the survival of patients with acute reparatory distress syndrome in association with COVID-19 is lower than that in association with other conditions [32,33].

In the unfavorable emergency context related to COVID-19, the French Society of Anesthesiology and Intensive Care has drawn up recommendations for the process of decision making about admission to the ICU [34]. These recommendations recall that the context does not justify renouncing the decision making principles of treatment limitations and discontinuation usually adopted by ICU practitioners. The goal is to preserve a medical decision based on deliberation while agreeing to allocate resources to those patients in whom interventions have the best chance of success. They recommend that the following five elements of decision making should be considered whenever possible:

- anticipating decision making to escape the emergency and guarantee the necessary time for deliberation is available to mature a decision;

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- collecting the patient's wishes directly, by relatives or through advance directives;
- maintaining collegiality with the reasoned opinion of at least one other doctor and conducting consultation with at least one member of the health care team;
- assessing the following aspects of the patient's state:
  - the patient's previous condition as reflected by age, comorbidities, the fragility evaluated by the Clinical Fragility Score [35], and the existence of a neurocognitive disorder,
  - current clinical severity as assessed by O_2 requirements of more than 6 L/min or respiratory distress, Glasgow score of less than 12 points, and systolic blood pressure of less than 90 mmHg,
  - the kinetics of aggravation of the patient's previous and current conditions;
- transparency with regular and repeated evaluations of the response to the therapies used, justification and traceability of decisions in the medical file, and provision of information to relatives.

Frailty is a recent geriatric concept that began to be discussed in the 1980s in North America. The Clinical Frailty Score, ranging from one to nine points, is an easy-to-use scale that includes various clinical items on cognition, mobility, physical fitness, and comorbidities. Several studies to date have reported its usefulness in predicting the functional prognosis of patients in the ICU [36,37]. Neuromuscular disorders may be incurable, but they are not untreatable, and the implications of different treatment decisions vary significantly. The neuromuscular specialist must be available to play a role in ensuring the fair provision of intensive care to patients with neuromuscular disorders.

In March 2020, a FILNEMUS multidisciplinary working group composed of neurologists, neuropediatricians, pneumologists, palliative care physicians, and ICU practitioners met to discuss choosing decision making criteria for the admission of neuromuscular patients to the ICU in the context of the COVID-19 pandemic.

The initial assessments of this meeting were as follows:

- many neuromuscular patients could be considered at high risk of complications from COVID-19 infection such as those with the presence of respiratory failure, cardiac involvment, long-term treatment with corticosteroids and/or immunosuppressive treatments, and comorbidities that may be associated with certain neuromuscular diseases (e.g., diabetes, obesity, and hypertension);
- the Clinical Frailty Score, used to assess the patient's previous condition, has never been validated in the context of neuromuscular diseases and could be a factor that overestimates a patient's prognostic severity;
- the large number of different neuromuscular disorders and their heterogeneity in terms of severity and prognosis make it difficult to assess the severity of the patient in an emergency context.

To promote the fairest possible collegial discussion for patients with neuromuscular diseases, the working group listed the neuromuscular diseases with a good prognosis, usually eligible for admission in ICU and, for other conditions not on this list, the positive criteria suggesting a good prognosis for admission to ICUs.

Conditions with a good prognosis for recovery include:

- autoimmune and congenital myasthenia;
- nondeficit metabolic myopathies, e.g., McArdle disease;
- inflammatory myopathies without severe systemic damage (in particular, pulmonary fibrosis);
- muscle channelopathies;
- most neuropathies, whether hereditary (Charcot–Marie–Tooth disease and FAP) or acquired (Guillain–Barré type, chronic inflammatory demyelinating polyneuropathy, and multifocal motor neuropathies with conduction blocks).

The positive criteria for a good prognosis for intensive care are:

- no major cardiac or respiratory damage and no major disability;
- for other patients, the criteria in favor of resuscitation are:
  - neuromuscular pathologies with slight progression,
  - respiratory functions being minimally impaired and stabilized,
  - mild and stabilized heart disease,
  - absence of severe thoracic deformities/severe contractures preventing ventral decubitus,
  - absence of multisystemic impairment and comorbidities,
  - preservation of autonomy for everyday life acts and/or social environment to supplement daily life tasks (e.g., presence of a family and caregivers).

It is noted that some patients with neuromuscular diseases are not autonomous in daily life but have a social environment that effectively compensates for this lack of autonomy.

As other procedures show a risk of generating aerosolization such as oxygen therapy, particularly at high-flow rates, it has been recommended that noninvasive ventilation (NIV) be avoided as much as possible to reduce the risk of contamination [38,39]. However, some patients with neuromuscular diseases use NIV chronically. If a patient has to use NIV, the use of a single circuit with a mask leak is contraindicated because of the aerosolization caused by the system. However, measures to adapt the circuit and the use of NIV make it possible to limit aerosolization and continue using NIV. Before starting or stopping NIV, the patient's mask must be in place, and personal protective equipment must be worn by caregivers [40]. The mask should be sealed as tightly as possible, involving a full face mask, perhaps with a temporary lowering of the pressure if the leakage is too great, and an anti-bacterial filter should be used at the ventilator outlet on the inspiratory circuit and after the mask [41]. Oxygen therapy can be added to the NIV if necessary. For patients with contraindications to resuscitation, we recommend that the local palliative care team be consulted.

Some neuromuscular patients have written advance directives, including whether or not they wish to be intubated in the event of respiratory distress. Emergency medical teams

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are reminded of the importance of checking with the patient's family and friends or referring physicians beforehand, if possible, to find out whether any advance directives have been drawn up. The regional services of the AFM-Telethon in their emergency kit updated during the COVID-19 period remind the patient of the importance of keeping these advance directives with them in the case of emergency.

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Disclosure of interest

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