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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French Neuromuscular Disease Network (FILNEMUS) guidance for care of neuromuscular patients during the COVID pandemic outbreak-19 of the French Neuromuscular Diseases Network (FILNEMUS)

Introduction:

COVID-19 outbreak has been responsible for challenges of medical practices in the context of COVID-19 since March 2020. It has significantly changed our current practice in neuromuscular disorders. These diseases constitute a group of very heterogeneous conditions, most often of genetic or autoimmune origin, affecting both children and adults, with a severity that varies widely from one individual to another. They include muscle disorders (muscular dystrophies, congenital myopathies, metabolic myopathies, inflammatory myopathies, muscle channelopathies, etc.), diseases of the neuromuscular junction (myasthenic syndromes either acquired or congenital) peripheral nerve disorders (dysimmune neuropathies, familial amyloid neuropathies, Charcot-Marie-Tooth disease, ...) and spinal muscular atrophies. (The guidance proposed here do not include amyotrophic lateral sclerosis).

In France, 40,000 to 50,000 patients are thought to suffer from neuromuscular diseases, excluding patients with amyotrophic lateral sclerosis. A significant number of patients are in a situation of great disability and may have cardiac and/or respiratory impairment.

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 the implementation of emergency measures and a degraded organization of neuromuscular reference centers. The

French Rare Health Care for Neuromuscular Diseases Network (FILNEMUS) has established guidance in order to homogenize the management of neuromuscular patients in this context with the aim of reducing the contamination of extremely fragile patients while avoiding the loss of chance linked to the interruption of essential treatment or follow-up.

In the uncertainty of the evolution of the epidemic, national measures have been proposed for a period of two months (at the time of writing of this manuscript for the period of March- April 2020) in order to homogenize care in France. However, it is difficult to make specific guidelines with regard to the heterogeneity of diseases and of the patients even for the same disease. Different scenarios taking into account regional specificities, particularly in terms of the severity of the epidemic, are then considered and will be the subject of subsequent guidance with view of offering optimal care to our patients, in accordance with to our public health responsibilities.

Information and communication to patients

Patients with neuromuscular diseases may have risk factors for developing severe forms of COVID-19 and prolonged intensive care may worsen their functional prognosis. It is therefore essential to develop a preventive approach. This involves easily intelligible information, the content of which must be adapted as knowledge progresses and widely disseminated. As patients are demanding for information and in order to homogenize the answers at the national level, a Frequently Asked Questions (FAQ) sheet has been compiled from the questions most frequently asked to the secretariats of the reference centers, the coordinator nurses and the regional offices of the AFM-Telethon, the French neuromuscular patient association. The main addressed topics were about the disease itself, its mode of transmission, the official national public health recommendations to prevent it, the behavior to adopt in case of symptoms, but also its consequences specific to the neuromuscular patient: management of steroids and immunosuppressor treatments, medical appointments, rehabilitation, ventilation, etc. The constant evolution of knowledge and official instructions have made it necessary to continuously update its content. The widest possible dissemination is necessary for this type of action to be effective. This FAQ is therefore available on the FILNEMUS website (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 within the reference centers or the regional offices of the AFM-Telethon. In addition to mission of disseminating information, these hotlines offer a link with patients and identify risk situations: loss of human aid, isolation, disruption of care, psychological distress, etc. The reference center

should therefore ensure that the patients have received relevant information and adequate equipment. Within the reference centers, these hotlines can be made up of paramedical staff working from home but also of residents redeployed from medical units whose activity has been reduced.

Appointment management and regular follow-up

In order to redeploy human and material resources in the fight against COVID-19, non-urgent appointments should be postponed or if possible, replaced by telemedicine (Lurie et al, 2018, Welscher, 2015). Tele-consultation was only possible by videoconference and after administrative steps that have slowed down its development (Decree No. 2010-1229 of 19 October 2010 on telemedicine) but the rules have been relaxed: when the patient does not have the necessary equipment to carry out videoconferencing, telecare activities can be carried out by phone. (Decree No. 2020-227 of 9 March 2020). A register of cancelled appointments must be kept by the reference center in order to reschedule them at a later date. Acts such as electroneuromyograms, muscle and nerve biopsies must be reserved for diagnostic emergencies: vasculitis, Guillain-Barré, myasthenia gravis, myositis...

Hospitalization should be reserved for emergencies, treatments that cannot be postponed (poorly balanced condition, relapse), check-ups for which the diagnostic delay may result in a loss of chance, cardio-respiratory assessments for which the delay could be detrimental to the patient. A register of cancelled hospitalizations must be kept by the centers in order to postpone them to a later date. A teleconsultation carried out by a doctor, a resident or even a coordinator nurse will enable to verify that hospitalization cancellation will not harm the patient and to determine the delay in rescheduling.

Management of the interruption of physiotherapy support

Maintaining joint flexibility, muscle strength and endurance are recommended in many neuromuscular diseases. They are ensured by regular in-house or office care by health professionals (Vry et al, 2016; Kang et al, 2015; Andrews et al, 2018; Apkon et al, 2018; Voet et al; 2019). The decree of 17 March 2020 regulating travel in the context of the fight against the spread of the COVID-19 virus led to the closure of private therapists' practices including physiotherapists, speech therapists and occupational therapists (Decree No. 2020-260 of 16 March 2020). A recommendation of the council of the order of physiotherapist masseurs calls

for "the maintenance of in-house physiotherapy of vulnerable patients for whom the cessation of care could lead to a major aggravation. Thus, the usual management of many patients is severely disrupted. During the period of outbreak, the High Council of Public Health recommends for the support of people with disabilities to develop activities allowing the continuity of the support when it is feasible at a distance (for example by tele-education). (HCSP. Epidemic in Covid-19: support for people with disabilities [Internet]. Paris: Haut Conseil de la Santé Publique; 2020 March).

A working group of FILNEMUS has set up various supports (illustrated sheets with links to online videos) for self-rehabilitation and exercises performed by caregivers or relatives. They are available on the FILNEMUS website (www.filnemus.fr) and free of copyright, they have been validated before distribution. Classified by theme (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 non-health professionals. It is reminded that these exercises have to be adapted to each situation under the control of the referring doctor and in close collaboration with the usual therapists. As most of the MNM reference centers and competence centers have maintained a significant teleconsultation activity, patients and families are encouraged to contact these departments for individualized support.

What to do about the treatment usually administered in hospitalization

In the epidemic context prior to the containment period, a national strategy has been adopted regarding treatments usually administered in daily or traditional hospitalization. This strategy has been developed for a period of one to two months and will need to be adapted thereafter according to the evolution of the epidemic in the regions.

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

The data entered in the French registry for Pompe disease allow us to estimate that 115 adult patients are treated with alglucosidase alfa (Myozyme®), and 20 are included in the clinical trials conducted by SANOFI (neoGAA and COMET trials) and AMICUS (PROPEL trial).

These enzyme replacement therapies, which are carried out in daily hospitalization, have been discontinued for most patients since the beginning of the epidemic in order to limit the risk of contamination in hospital, especially since most patients with Pompe disease have respiratory insufficiency related to diaphragmatic involvement.

It is likely that stopping enzyme replacement therapy over a period of one to three months will probably not lead to significant worsening of the disease, although there are few data available in the medical literature to estimate the risk of worsening the disease after a relatively short interruption of treatment. A Swiss study of 7 patients whose treatment was interrupted for economic reasons (Hundsberger et *al.,* 2014) showed a clear deterioration of motor and respiratory functions when treatment was stopped, and no complete recovery after resumption of treatment (Scheidegger et *al.,* 2018). However, in this study the duration of treatment interruption was greater than 9 months in 6/7 patients, and there was no clinically significant worsening in the patient with the shortest duration of interruption of 3 months.

For infantile form of Pompe disease, the experts confirm the benefit of the early treatment for cardiac and muscular function and the necessity to initiate or to continue the treatment as usual.

Assuming that many of the daily in-patient units would remain closed for several weeks or months, the possibility of home infusions could be considered to limit the risk of contamination of patients in 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 will be resumed, a motor and respiratory functions evaluation should be carried out, with pulmonary function and 6-minute walking tests to assess the impact of this suspension of infusions.

- **Intrathecal injections of @*nusinersen* for spinal muscular atrophies**

Initiation of nusinersen represents a therapeutic emergency for children with type 1 or type 2 SMA. The indication should be maintained because of the functional and vital consequences of delaying treatment. For adolescent and adults with type II SMA and type III 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 for several months (Rigo et al, 2014). In addition, there appears to be a cumulative phenomenon as intrathecal concentrations of nusinersen dosed prior to injection increase on average from 1.68 ng/ml 15 days after the first injection to 7.46 ng/ml 4 months after the 6th injection (Finkel et al, 2016). It is recommended to try as much as possible to continue intrathecal injections of type 1 and of young children type 2. For adolescent and adult, injections could be delayed from 1 to 4 months depending of the evolution of the disease. Furthermore, for patients with arthrodesis,

CT-guided injections are no longer available in most University Hospitals due to the reorganization of services.

Several dose catch-up schemes are possible:

- Rapid catch-up for the most progressive forms
    - Dose delayed by less than 3 months: inject as soon as possible and restart the schedule at the 8th month initially planned.
    - Dose delayed by 4 months: inject two doses 15 days apart.
- Slow catch-up for the less progressive forms
    - Delayed dose of less than 3 months: injection as soon as possible and continued every 4 months.
    - Dose delayed by 4 months: restart the treatment regimen every 4 months

- **Gene therapy for SMA (@Zolgensma)**

Since June 2019, Zolgensma treatment is allowed in France for SMA type 1 aged under 2 years old with a specific national procedure (ATU) after evaluation of every case by a national expert committee and the agreement of the National Health Assurance (ANSM) For the moment, 12 children were treated. This committee decided to continue the same process and the administration of the treatment is performed in ICU of identified pediatric hospital with the detection of COVID infection (PCR) before treatment. After 3 days, they come back at home with closely medical and biological evaluation.

- **Treatment by patisiran for familial amyloid neuropathies (FAP)**

It is recommended to interrupt patisiran infusions in hospital to avoid exposing these patients to the risk of COVID-19 contamination in the hospital environment as these patients are considered to be at high risk of serious complications (frequent age >70 years, underlying cardiomyopathy, ...) with a relay carried out by infusions performed at home. The time to organize the relay, the interruption of one to 2 cures should not pose any particular problem.

The Phase 2 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 (Suhr OB et al, 2015). In the Phase 2 and Phase 3 extension clinical trials (APOLLO), the interruption of 1 to 2 infusions of patisiran, authorized in the protocol, involved several patients without having a negative impact on the final results

(Adams D et al, NEJM 2018). In addition, no anti-SIRNA antibodies were observed in any of the treated patients.

- **Intravenous immunoglobulins**

Treatment of dysimmune neuropathies (chronic polyradiculoneuropathy, Lewis-Sumner disease, multifocal motor neuropathy with conduction blocks, etc.) is most often based on the iterative administrations of either intravenous or subcutaneous immunoglobulins, carried out in hospital or at home, the benefit of which has been widely demonstrated (Farmakidis et al, 2020). Interruption of the therapy may result in the re-exacerbation of the symptoms of neuropathy. Home therapy when available is a very good alternative but unfortunately this latter is not always available for many, Moreover, this in view of enhanced staff shortage and is unsuitable for treatment initiation. It is not recommended to switch to oral corticosteroids (Farmakidis et al, 2020), since this treatment is not recommended during SARS-CoV-2 infections and may worsen the disease in dysimmune motor neuropathies (Russell et al, 2020). In autoimmune myasthenia gravis, intravenous immunoglobulins are also used, either during an outbreak or recurrently (Nguyen-Cao et al, 2019). In the latter case, the recommendation for home therapy was also made, as for dysimmune neuropathies, unless the severity of the relapse warranted in-patient monitoring.

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 their treatments in the absence of any manifestation suggestive of COVID-19. It should be reminded that stopping treatment may lead to relapse of the disease. Particular attention should be paid to steroids therapy, since 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 to initiate immunosuppressant treatment (azathioprine, methotrexate, mycophenolate mofetil…) in order to control a severe inflammatory pathology. This will 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 in order to prevent the patient from being contaminated by the home care nurse. The

decision depends on the reason for choosing the injectable form. If it was a reason of efficacy, then optimal disease control is necessary. If it was a problem of tolerance, it is difficult to reverse it, possibly with a dose splitting trial.

As for biotherapies (rituximab and equivalents ...), it is justified to maintain them if they are effective and well tolerated, in order to avoid the potential occurrence of a relapse, with strict application of public health recommendations. For the initiation of a biomedication, a decision 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 (International MG/COVID-19 Working Group et al, 2020).

In 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 in respiratory syndromes such as SARS or MERS, without worsening or even improving mortality rates (Stockman et al, 2006, Arabi et al, 2018). Data about other treatments, the data remain very sparse. Nevertheless, the first results of the Italian experience are demonstrative. Out of a cohort of 320 chronic inflammatory rheumatic diseases treated by biotherapies or JAK inhibitors, 8 patients had a COVID-19 infection either confirmed or strongly suspected (Monti et al, 2020). In all cases, the authors report that they suspended treatment during the infection. Treatment was resumed 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, resumption of immunosuppressant therapy should be discussed one to two 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 it is decided to maintain immunosuppression, 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 (Zhang et al, 2020).

Conduct of Therapeutic Trials

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

What to do about cardiac treatment

Some patients have been tempted to stop their Angiotensin Converting Enzyme (ACE) inhibitors or Angiotensin II Receptor Blockers (ARB) treatments in view of the information (mostly transmitted on social media) that these drugs may increase both the risk of infection and the severity of SARS-CoV2. The concern arises from the observation that this virus binds to ACE2 to infect cells, and ACE2 levels are increased in patients treated with ACE inhibitors and ARBs. However, there is currently no data proving a causal relationship between ACE2 activity and SARS-CoV2 associated mortality (Vaduganathan et al, 2020). Some findings from preclinical studies even suggest a possible protective role of ARB in SARS-CoV2 associated lung injury (Kuba et al, 2005). It has been well shown that abrupt withdrawal of ACE inhibitors or ARBs treatments in high-risk patients, including those who have cardiac dysfunction, may result in clinical instability and adverse health outcomes.

As a result, patients who are at risk for, being evaluated for, or with Covid-19 should continue their usual cardiac treatments, including ACE inhibitors and ARBs (https://www.escardio.org/Councils/Council-on-Hypertension-(CHT)/News/position-statement-of-the-esc-council-on-hypertension-on-ace-inhibitors-and-ang).

Particularities of neuromuscular patients with respect to treatments prescribed for COVID-19

Numerous therapeutic trials have been initiated in the context of COVID infections. Although these treatments are promising, their effectiveness needs to be confirmed in a larger population. 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. A large randomized European study (DISCOVERY) evaluating 4 treatments was started in March 2020 in COVID. The 4 types of treatments are: remdesivir, lopinavir in combination with ritonavir, the latter being associated or not with beta interferon, and hydroxychloroquine (ClinicalTrials.gov Identifier: NCT04315948).

In the context of other neuromuscular diseases, particular attention must be paid to two treatments, hydroxycholoroquine and azithromycin, which have been proposed for COVID-19 infections (Gautret et al, 2020).

A retrospective study of 127 patients with autoimmune myasthenia gravis showed a worsening of the disease after taking treatments such as azithromycin (odd ratio: 1.42), fluoroquinolone (odd ratio: 0.89) and beta-blockers (2.70). These treatments are also contraindicated in congenital myasthenia gravis (Abicht et al, 2016). 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 worsening or flare-up of myasthenia gravis usually occurs within days after administration of the contraindicated antibiotic treatment, but flare-ups may occur later up to 29 days after initiation of treatment (Gummi et al, 2019). In the absence of demonstrated benefit from (hydroxy-chloroquine and given that an exacerbation of myasthenia gravis may be caused by these molecules, the FILNEMUS network recommends 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 paid to the fact that the primary cardiac effect of hydroxycloroquine is QT prolongation which may lead to cardiac arrest secondary to cardiac arrythmia. Among neuromuscular diseases, the one that should most encourage caution is. This risk is increased when combined with azithromycin, which has the same cardiac effects. The decision to initiate or continue this treatment can only be based on an individual benefit-risk assessment taking into account 1) QTc measurements and the occurrence of cardiac arrythmia, 2) the effectiveness of the treatment on COVID infection, and 3) the presence of QT modulating factors (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 chronic treatments.

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 raised major concerns about the capacity of the various health systems to cope with the increased medical needs, particularly in terms of resuscitation beds. In France, the medical community, and particularly ICU practitioners, have raised the possibility of having to make difficult choices and prioritize patients in an emergency regarding access to resuscitation. The risk factors for developing severe COVID-19 infection seem to be age and the association of co-morbidities. Recent papers also report that the survival of patients with acute reparatory distress syndrome in association with COVID-19 is lower compared to that in association with other conditions (Arentz et al, 2020; Wu et al, 2020).

In this unfavorable emergency context related to COVID-19, the French Society of anesthesiology and Intensive Care (SFAR) has drawn up recommendations on the process for decisions on admission to intensive care unit (ICU) (see ref). These recommendations recall that the context does not justify renouncing the decision-making principles of treatment limitations and discontinuation (LAT) usually taken by ICU practitioners. The goal is to preserve a medical decision based on deliberation, while agreeing to allocate resources to those patients for whom interventions have the best chance of success. These texts recommend that five elements of decision making should be taken into account whenever possible:

- The anticipation of decisions in order to get out of the emergency and guarantee the necessary time for deliberation to mature a decision.
- Collection of patient's wishes directly, by relatives or through advance directives
- Maintaining collegiality with the reasoned opinion of at least one other doctor, and consultation with at least one member of the health care team.
- The decision-making elements are:
    - the patient's previous condition reflected by age, co-morbidities, the fragility evaluated by the Clinical Fragility Score (Rockwood, 2005), the existence of a neurocognitive disorder
    - Current clinical severity as assessed by O2 requirements >6L/min or respiratory distress, Glasgow score<12, systolic blood pressure <90mmHg
    - The kinetics of aggravation of his previous and current condition.
- Transparency with regular and repeated evaluation of the response to the therapies used, justification and traceability of decisions in the medical file and information for relatives.

Frailty is a recent geriatric concept that developed in the 1980s in North America. The Clinical Frailty Score (CFS), ranging from 1 to 9, is an easy to use scale that includes various clinical items on cognition, mobility, physical fitness and co-morbidities. Several studies have reported its usefulness in predicting the functional prognosis of patients in ICU (Bagshaw et al, 2015; Nakajima et al, 2019; Fernando et al, 2019). Neuromuscular disorders may be incurable, but they are not untreatable, and the implications for treatment decisions are very different. The

neuromuscular specialist must be available to play a role in ensuring fair provision of intensive care to NMD patients.

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

The initial assessments were that:

1) Many neuromuscular patients could be considered at high risk of complications from COVID-19 infection: presence of respiratory failure, heart disease, long-term treatment with corticosteroids and/or immunosuppressive treatments, co-morbidities that may be associated with certain neuromuscular pathologies (diabetes, obesity, hypertension).

2) The CFS, used by to assess the patient's previous condition, has never been validated in neuromuscular diseases and could be a factor that overestimates patient's prognostic severity.

3) 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.

In order 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 resuscitation and, for other conditions not on this list, the positive criteria for a good prognosis for admission to critical care and/or resuscitation units.

Conditions with a good prognosis for recovery are:

- Autoimmune and congenital myasthenia
- Non-deficit metabolic myopathies: e.g. Mc Ardle disease
- Inflammatory myopathies without severe systemic damage (in particular, pulmonary fibrosis)
- Muscle channelopathies
- Most neuropathies, whether hereditary (Charcot Marie Tooth disease, FAP) or acquired (Guillain-Barré type, Chronic inflammatory polyradiculoneuropathies, multifocal motor neuropathies with conduction blocks).

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

- Patients without cardiac or respiratory damage and without major disability
- For other patients, the criteria in favor are:
    - Neuromuscular pathologies with slight progression
    - Respiratory functions minimally impaired and stabilized
    - Mild and stabilized heart disease
    - Absence of severe thoracic deformities / severe contractures preventing ventral decubitus.
    - Absence of multisystemic impairment and co-morbidities
    - Preservation of autonomy for everyday life acts and/or social environment to supplement daily life tasks: presence of a family, caregivers, etc.

It is pointed out 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 at risk of generating aerosolization such as oxygen therapy, particularly at high flow rates, it has been recommended that Non-Invasive Ventilation (NIV) should be avoided as much as possible in order to reduce the risk of contamination (Cheung et al, 2020; Hui et al, 2019). 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 nebulization caused by the system. However, measures to adapt the circuit and the use of NIV make it possible to limit nebulization and continue using NIV. Before starting or stopping NIV, patient's mask must be in place and personal protective equipment must be worn by caregivers (https://www.srlf.org/wp-content/uploads/2020/03/Recommandations-dexperts-COVID-9-mars-2020.pdf). The mask should be sealed as tightly as possible, including a mouth mask, possibly with a temporary pressure relief if leakage is too great, and an infection filter should be used at the ventilator outlet on the inspiratory circuit and after the mask (https://www.sfrms-sommeil.org/wp-content/uploads/2020/03/LES-PROCEDURES-DU-GAVO2-ProtectionVirale2020-MAJ14mars2020.pdf). Oxygen therapy can be added to the NIV if necessary. For patients with contraindications to resuscitation, we recommended that the local palliative care team should 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 are reminded of the importance of checking with the patient's family and friends or referring physicians beforehand, if possible, to find out if there are any advance directives that 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 case of emergency.

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