Clinical Guidance on COVID- 19 Vaccines for People with Significant Neuromuscular Conditions Who Require Respiratory Support

This guidance is intended for health-care providers and is based on known evidence as of April 18, 2023.

Patients with neuromuscular conditions with significant respiratory muscle weakness are at increased risk of hospitalization and mortality from COVID-19.[1] This includes individuals with significant diseases of the neurologic system including the brain, spinal cord, motor nerves and muscles who, because of their condition require respiratory support in the form of home ventilation or bilevel positive airway pressure in order to function in daily life.[2,3]

This includes individuals requiring respiratory support with the following conditions:

- Motor neuron disease
- Muscular dystrophy
- Peripheral neuropathy including Guillain Barre Syndrome, Charcot-Marie Tooth disease, critical illness neuropathy
- Myopathies including congenital myopathies, myofibrillar myopathies, metabolic myopathies, critical illness myopathy
- Other neuromuscular conditions where breathing muscles are severely impacted due to their conditions
- While people with spinal cord injury are not considered to be at increased risk of getting infected with the COVID-19 virus,[4] those with a spinal cord injury requiring ventilatory support have the same risk factors as other conditions requiring respiratory support mentioned above, thus the clinical judgment is that their risks are similarly high.

Is COVID-19 immunization recommended for patients with neuromuscular conditions who require respiratory support?

COVID-19 immunization should be encouraged for patients with neuromuscular conditions requiring respiratory support and is not contraindicated, including those who have had COVID-19 infection. This recommendation is based on the following factors:

- Patients with neuromuscular conditions who require respiratory support at baseline are at extremely high risk for morbidity and mortality if they are infected with COVID-19; many would not be able to be extubated if intubation was required.

- Weakness of respiratory muscles in individuals with neuromuscular disorders may result in impaired ability to take a deep breath, impaired cough reflex, and ineffective airway clearance of secretions predisposing to atelectasis and lung infection.[2] Acute respiratory failure may rapidly evolve in patients with chronic respiratory failure secondary to neuromuscular weakness. Risks include prolonged invasive ventilation, deterioration of respiratory or skeletal muscle function or death.[1]

While data specific to the safety and efficacy of COVID-19 vaccines for people with neuromuscular disorders is currently limited, there are data to suggest that the currently available COVID-19 vaccines have efficacy.[5] The authors of this guidance agree that the benefits of vaccine-induced immunity against COVID-19 for this population outweigh any theoretical risks of immunization.

Is COVID-19 immunization efficacious and safe for patients with neuromuscular conditions who require respiratory support?

Patients with neuromuscular disease requiring respiratory support were not specifically included in the COVID-19 vaccine trials; therefore, efficacy in this population is unknown.[6-11] However, there is no reason to believe the vaccine will be less efficacious in patients with neuromuscular disease requiring respiratory support than in the population studied in the clinical trials. Patients with chronic pulmonary disease comprised 7.8% of patients in the Pfizer-BioNTech vaccine trial and patients with hemiplegia and paraplegia comprised 0.1% of patients in the trial.[12]

The Food and Drug Administration (FDA) have issued a for the Janssen COVID-19 vaccine about the increased risk of developing Guillain-Barré syndrome (GBS) in the 42 days after vaccination.[13] The GBS/CIDP Foundation recommends that patients who have developed their disease within 6 weeks of receiving a COVID-19 vaccination, they should make an informed consent after discussing the risks versus benefits with their healthcare professional about receiving a second dose of vaccine that is of a different type, preferably mRNA, as per the NACI guidance.[14]

Patients with Duchenne's Muscular Dystrophy (DMD) who require respiratory support and who are receiving deflazacort or prednisone will require additional counseling on efficacy and timing of their vaccine with their treatment, as deflazacort and prednisone are immunosuppressing/immunomodulating. There is limited evidence about the efficacy of the Pfizer BioNTech and Moderna vaccines in people who are immunocompromised due to treatment, as immunocompromised patients were not included in the trials. It is unknown if the currently available COVID-19 vaccines are efficacious in those who take immunosuppressants compared to those who are not considered immunosuppressed.

- It is possible that, because of their immunosuppression from treatment, these patients will have a blunted immune response to the vaccine. Because of their increased risk to COVID-19, the vaccine is recommended for patients with neuromuscular conditions who are immunocompromised, but these patients should be informed that they may have a diminished immune response to any of the authorized COVID-19 vaccines.[12,15] As per NACI, safety data in immunocompromised individuals, including those receiving immunosuppressive therapy, were available from observational studies in people who were taking immunosuppressive therapies. The frequency and severity of adverse events following vaccination with an mRNA COVID-19 vaccine were comparable to that

of non-immunocompromised individuals in these studies and what was reported in clinical trials. Safety data in these populations following vaccination with a viral vector vaccine is not available.

- Health-care providers caring for DMD patients being treated with deflazacort or prednisone can refer to the clinical guidance for patients with neuromuscular receiving immunosuppressing/immunomodulating therapy.

Are there any specific contraindications or exceptions for patients with neuromuscular conditions who require respiratory support?

Individuals who have had a severe allergic reaction to an ingredient of one type of COVID-19 vaccine are still able to receive future doses of the other type of vaccine.[16] BCCDC has a list of the individual components and their purpose in the vaccines. For a complete list of components in the vaccine, consult the vaccine monographs found at: www.bccdc.ca/health-info/diseases-conditions/covid-19/covid-19-vaccine/vaccines-for-covid-19.

For individuals with a history of anaphylactic reaction to a previous dose of an mRNA COVID-19 vaccine, re-vaccination (i.e., administration of a subsequent dose in the series when indicated) may be offered with the same vaccine or the same mRNA platform if a risk assessment deems that the benefits outweigh the potential risks for the individual and if informed consent is provided. Prior to revaccination, consultation with an allergist or another appropriate physician (e.g., Medical Health Officer) is advised. If re-vaccination is going ahead, vaccine administration should be done in a controlled setting with expertise and equipment to manage anaphylaxis, with an extended period of observation of at least 30 minutes after re-vaccination.

Health Canada continues to monitor any adverse events following immunization through their post-authorization surveillance process.

COVID-19 vaccines can be given concomitantly with, or any time before or after any other live or inactivated vaccine.[17-20]

Are there specific recommendations or considerations for safe and/or most effective vaccine administration?

Individuals with muscle disease may not have adequate deltoid muscle mass, in which case the anterolateral thigh can be used to administer the vaccine.[21]

Otherwise, there are no other specific recommendations that pertain to this population unless they have comorbidities requiring special care, such as being treated with immunosuppressive or immunomodulating therapy, in which case health-care providers can refer to clinical guidance for people with autoimmune neuromuscular disorders receiving immunosuppressive/immunomodulating therapy.

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