

## Webinar 6: Duchenne Muscular Dystrophy and COVID vaccines

WDO webinar, Saturday February 28 ([link to recording](#))

*During the WDO Member Meeting on February 28, the World Duchenne Organization invited Dr. Erik Niks to present a webinar about Duchenne Muscular Dystrophy in relation to the COVID-19 vaccines. Dr. Niks is a pediatric and adult neurologist in Leiden University medical Center (NL). He discusses questions often received from both families and clinicians, and proposes guidance on getting your vaccination related to being on steroids.*

**Important notice: Changes in your steroid regimen should always be discussed with the one prescribing it, which is often a clinician.**

### What is the best way to prevent a COVID infection?

The viral spread basically takes place through coughing and sneezing. For this reason, we ask people to stay indoors and to avoid contact as much as possible. The transport is via droplets by the air, however these droplets can also contact surfaces. That's why it's important to keep good hand hygiene. You either get infected through airborne transmission or by contact with nose, mouth or throat transmission. Mild infections basically resemble symptoms of the common cold: you get a fever and start coughing. More specific for Corona is that you also can have a loss of smell and taste. This can stay for quite some months after the infection. Many people have been complaining about pain in the eyes or muscles. The last one is also something you see in the common cold. Another symptom of the common cold is gastrointestinal symptoms like vomiting and diarrhoea.

### Why is a COVID infection so dangerous?

Normally COVID is a self-limiting disease that lasts a few days of weeks. A small proportion of patients get a severe infection and are submitted to hospitals. Some end up in the ICU in the case of a large-scale inflammation in the lungs and other organs. In this case it is not the virus anymore, but the reaction of your immune system in response to the viral infection that is now affecting lungs and other organs.

### How do most COVID vaccines work?

The virus looks like a round particle with protein spikes at the border. The spike protein is one of the key players for the virus to enter the body. If you come in contact with the Coronavirus, the spike protein recognizes certain receptors within the body, leading to a spread, and in some severe cases to large inflammations. There are many vaccines in development. Some of them already gained approval by the FDA and/or the EMA, and some are still ongoing. Nearly all the vaccines are targeting a similar spike protein.

### Why is there an age limitation on the vaccination?

It's important to realise that all of the trials have been performed in adults. The AstraZeneca vaccine has been tested in people 18 years and older. Same is the case for the Moderna vaccine. Only the Pfizer RNA-based vaccine is tested in participants 16 years and older. This has to do with the fact that the regulatory approval is given for use in people from either 16 year onwards or 18 years onwards. The Moderna vaccine has a trial ongoing with children between 12 and 18 years of age.

## How can the vaccine trigger an immune response?

The spike protein has a DNA code: a gene encodes for this protein. This gene is taken out of the virus and is wrapped into an adenovirus. This is another type of virus. Some of them are involved in a common cold, so our body is very capable of taking care of these virus particles in the body cells. In case of a COVID infection, our cells start to produce the spike proteins because we now have the genetic code to make this protein. This protein is again expressed on the surface of the body cells. Because it's expressed on the body and on the surface, our immune system (our white blood cells) are able to recognize this protein and produce antibodies. This is the essence of the immune response. Of course, this takes time, but if you have this vaccination done and the antibodies are ready, you are much more capable of responding quickly if you come into contact with the actual Corona virus.

## Do people with Duchenne have a bigger risk to get COVID?

It's common sense to think that people affected by Duchenne are specifically sensitive to a Corona infection. Older children and young adults with DMD develop respiratory weakness and need respiratory support. However, although we are a year into the pandemic and Duchenne patients do not seem to be specifically sensitive to a Corona infection. At least, not to the development of the severe immune response. This might have to do with the fact that paediatricians do not see children with severe symptoms of COVID. They do develop symptoms, but in general they are very mild and do not lead to ICU admissions. We also know that families with DMD are very strict in following all hygiene guidelines. Another hypothesis is that the use of steroids may play a role. Steroids dampen the inflammatory response. Dexamethasone or another type of steroid is now used in the ICUs to treat severe Corona infection. It might be that the use of steroids is also protecting you in some way.

## Are the vaccines safe for people with Duchenne?

A question we also often receive is whether vaccination is safe in people with Duchenne. From all experts I've spoken, they all agree that vaccination is safe. The general recommendation is to provide vaccination in patients with other types of immunosuppressive therapies such as rheumatology diseases and other neurological diseases where people get much more immunosuppressive therapies than the steroid treatment that is given in Duchenne. The general recommendation is also to provide vaccination to those patients, so it's considered to be safe.

## Does the vaccine interfere with other DMD medication?

Many people with Duchenne use other types of medication, such as cardiac medication or antibiotics (sometimes as maintenance). We think there's no reason to either suspend or change the current medication that you use, either when there is a COVID infection or prior or after the vaccine is provided.

## Which COVID vaccine is best for people with Duchenne?

There are many statistics available on the percentage of protection against the Corona virus. The most important thing to stress is that all vaccines so far are tested to protect against the severe form of a corona infection: the vicious inflammatory response. This is an efficacy of all vaccines, so there is no reason to avoid a specific type of vaccine.

### Is being on steroid a reason to not get vaccinated?

We also received this question from doctors, because people with Duchenne get treated with steroids for many years. Vaccination is considered to be safe with any type of immunosuppressive therapy. Being on steroids is not a reason to not get the vaccinations. However, you need to have an immune response to build up your protection against the Corona virus. It could be that the vaccination is a little bit more effective if you're not using steroids. This is something that we are likely to collect scientific data on, but this is a reasonable hypothesis that your immune response to the vaccine is a little bit more effective if you do not use steroids.

### Is it smart to decline my vaccine appointment?

As in some countries planning for vaccinations are still under development and it will be challenging to get one, any delay of vaccination is not preferable. Sometimes it can be (too) difficult to change an appointment. We recommend to get the vaccine as soon as possible, taking earlier points into consideration.

### Do I change my scheme when I'm on 10 days on, 10 days off steroids?

With the 10 days on, 10 days off scheme it might occur that both vaccination dates are in a period where the patient is off treatment. If the person does well in the periods without steroids and is perceived to not be steroid dependent, you can skip one treatment cycle. This means the vaccination date will fall in the time when the person is off steroids. Another thing you can do is to shorten the period for a bit. For example: you shorten the treatment from 8 to 3 days. There is no difficulty in shortening one cycle of treatment. On an individual basis, it is best to discuss this with the person who prescribed the steroids.

### Do I change my scheme when I'm on daily or daily alternating steroids?

In the case of daily steroids, or one day on, one day off, you cannot presume you are not dependent on the steroids. Our urgent recommendation here is to not change the schedule, and get the vaccinations as they are originally planned.

### If I get the vaccine, will I be able to join clinical trials?

Right now, vaccinations are not a problem for future participation. If you look at the currently developed gene therapies, they use an adeno associated virus or AAV. Although this name resembles the adenovirus (AV) it's a completely different type of virus. The AAV does not have a spike protein that the vaccines target. So, if you get the vaccination now, it's not a problem to have gene therapy trials using associated viruses. We have seen a few protocols in which they include the possibility of joining when you received the vaccine. Next to this, most clinical trials are conducted in children, so below the age of 18. Most vaccines are approved for use in people older than 18 years.

## Summary

- We don't have any experience that people with Duchenne MD are specifically vulnerable to the severe form of COVID.
- It is important to get vaccinated, as we still consider it the only protection against the severe complications of the Coronavirus.
- Vaccination is currently planned for people 16 years and older (18 in some countries)
- Vaccination is safe
- Vaccination can be given with current medication including steroids
- Vaccination may be more effective without steroids
  - ONLY the 8-10 days on/off scheme can be adapted to the vaccination
  - DO NOT change daily or alternate day dosing