

Webinar 2: COVID-19 and Duchenne and Becker muscular dystrophy

Script of WDO webinar, Saturday March 21 – 4pm CET

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What we know about COVID-19

At the last webinar, we had Professor Mercuri, Professor Muntoni, Professor Finder and professor Goemans to share their opinions on questions we gathered from the community. You can find the [recording](#) together with the [report](#) on our website. To summarise these insights, we created a list of [15 things we know so far](#) about COVID19 in relation to Duchenne and Becker muscular dystrophy.

Clinical trials

Until now, there was a lot of uncertainty with regard to how EMA and FDA would deal with issues around clinical trials. Both agencies have now shared documents to provide guidance on how to manage the conduct of clinical trials in COVID-19 times. At the same time, sponsors and companies are sending us their plans and schedules, which can be found on the [WDO live feed](#).

Dr. Jarod Wong – endocrinologist at Glasgow University

What is the effect of steroids on the immune system?

Steroids prescribed for DMD may have **some impact on lowering the immune system**. Hence, routinely flu vaccination is recommended. People taking steroids have been identified as an at-risk group in the current climate by some governments. However, we do not commonly see severe, unusual and serious infections in people with DMD on steroids.

What is the effect of steroids and COVID-19 infection in DMD?

At the moment, we are not aware of any cases of people with DMD and COVID19 infection. It is theoretically possible that if infected, the infection may be more severe. However, **we simply do not know**. In some countries, any person on long term steroids has been classified as at-risk and recommended to isolate for a longer period of time, for instance 12 weeks.

Should I stop steroids in this instance then?

No, this should not happen and is not possible. One issue with anyone taking steroids for a prolonged period i.e. longer than a few months is that the adrenal glands, which make steroids naturally, are suppressed (ADRENAL SUPPRESSION LEADING TO ADRENAL INSUFFICIENCY). Even if we do want to stop steroids, a slow plan of gradual reduction over several months is essential.

To cope with severe infection, extra steroids are needed - stress dosing. Otherwise, the person could become very ill and be in an adrenal crisis. One possibility of more severe infection (of all kinds) in people with adrenal suppression from taking steroids may be that steroid management during the illness is not adequate.

Is there anything extra you need to do if on steroids during (COVID) illness?

Regardless of the type of infection in a person with DMD taking steroids, if the person has vomiting and/or diarrhoea, steroids should be given in another form. If there is access to steroids in the form of hydrocortisone injection at home, this needs to be given and then presented to the hospital.

In some people with DMD on steroids (especially older boys or men on lower doses of steroids), there **may be a need to increase the dose of oral steroid** during mild to moderate illness, which includes fever. It would be worth checking with your neuromuscular team if this is needed. In the majority of cases, this may not be necessary. Some teams have been advising all their patients to do so to be on the safe side. Generally, this should be for a period of 48 hours but may be longer if the person is sick. Currently, if the symptoms might be COVID related and do not resolve within 48 hours, generally the advice is to contact the relevant places for COVID advice, for instance the national hotline.

For those on intermittent steroids, a steroid plan should be in place with information on what to do if the person with DMD is unwell during the days off steroids.

What do you need to consider if admitted to hospital?

It is very important that if a person with DMD is admitted to hospital for any reason you remind the doctors that steroids cannot be stopped. The key words I recommend you to use with your treating medical team are as below. It is a very important issue and should not be the LAST thing we think about:

L-ongterm steroid treatment

A-drenal suppression

S-teroid dependent

T-reat with extra steroids when unwell

If the person with DMD on steroids is very unwell, especially in intensive care, steroids should be given via drip.

Where can I or my treating doctor find more information?

Most centres and hospitals have protocols to manage adrenal suppression during illness. If the treating doctor is not familiar with this issue or with DMD, the **PJ Nicholoff Protocol** has very useful and detailed information, including treatment doses of steroids. The protocol was published in a scientific journal in 2017, which the doctor will be able to access. A patient friendly summary is also available online.

[Published journal article for medical professionals](#)

[Patient summary](#)

My son was supposed to start steroids now. Should I wait until the pandemic is over?

In the current climate, we need to **consider what is practical**. Pharmacies in most countries are extremely busy and may also not dispense new medication. There are also tests that are needed prior to starting steroids; and follow-up after starting steroids is needed. So, being realistic, starting steroids currently is not likely to happen.

How do I convince my doctor/hospital to listen to me?

For doctors who are not familiar with DMD related issues, you could point them towards online information on the condition. For instance, as mentioned, regarding steroid management during illness for DMD, you can point them to the PJ Nicholoff Protocol. It may also be helpful if you have copies of your clinic letters to show doctors who are not familiar with the condition. Finally, you could also strongly encourage the treating doctor to contact your neuromuscular team.

Prof Dr. Annamaria De Luca – pharmacologist in Italy

Any immune boosting drugs, vitamins, supplements to fight/lighten infection?

There are no dietary supplements that can help in the case of COVID-19. All vitamins, minerals and other components that allow proper function of our immune system are in healthy food. The advice is to **maintain a healthy diet that is properly varied** with lots of fruit, vegetables, fish, meat and fibre. In some specific vitamin deficiency cases, such as vitamin D or calcium in Duchenne, extra supplementation is important and needs to be maintained.

A natural compound is not by definition safe and can even cause harm. This may occur at high dosages, or there might be harmful interactions with drugs used in the Standards of Care or for other necessary therapies (i.e. during the COVID-19). Supplements that are not purchased in a pharmacy or from authorised suppliers can contain impurities and contaminants that can have serious consequences for our health. So, **self-medication without the control of health specialists, and acquisition on the internet are not recommended.**

The same goes for melatonin, which is claimed to have a wide variety of effects, including being antioxidant and anti-inflammatory. This endogenous compound controls circadian rhythm and is mostly used for regulating sleep. There is no strong scientific evidence for the relationship between melatonin levels and the severity of an infection, so use of this is not encouraged without control of doctors for the reasons detailed above.

Is it true steroids might have a positive effect on COVID-19?

It has been proposed that low dosage of steroids can be useful in a so-called cytokine storm. This is a severe phenomenon that might occur in COVID-19 patients in an advanced stage of pneumonia. Normally, our immune system can combat the infection, however at a certain stage there could be an excessive discharge of the virus from the infected cells, leading to a massive production of cytokines. China proposed that low doses of glucocorticoids such as alpha methyl prednisolone can help reduce the storm without causing immunosuppression. There are clinical trials ongoing in COVID-19 patients, but there is some debate about the real usefulness of steroids in this condition, especially in patients already on steroids.

With the information and data we have, **it's important not to stop steroids unless specifically indicated.** This is also valid for other Standards of Care in patients, i.e. those receiving treatment with ACE inhibitors. It's best to maintain drugs that are effective in controlling cardiovascular function, as evidence of the potential risk of ACE inhibitors are few and controversial.

What is the best drug to treat fever in case of COVID-19?

There are many reports about the preferential use of paracetamol, rather than ibuprofen and other non-steroidal anti-inflammatory drugs (NSAIDs) in case of COVID-19. NSAIDs have been suspected to cause an extensive anti-inflammatory reaction that might not be useful, if not dangerous, in case of a virus infection. This is, however not confirmed, and recently the European Medicines Agency said

that patients with COVID-19 can use anti-inflammatory drugs according to what the doctor decides in relation to the drug and the risk benefit ratio.

For Duchenne and Becker patients, this can be different. Many non-steroidal anti-inflammatory drugs are not advised because of the concomitant use of steroids. In this setting, **paracetamol is the first choice of drug to treat fever** and has to be used at low dosage for the shortest time possible.

I would like to underline that fever is an important symptom for the diagnosis of COVID-19, so if fever occurs, a doctor has to be able to make a proper diagnosis.

Is chloroquine useful for treating the infection? Is it safe in DMD patients?

It's important to say that at the moment there is no approved therapy for COVID-19. While research is aimed at finding specific drugs against the coronavirus, a lot of effort is being put into searching for effective drugs among those already available. It is a dynamic field and we are learning from what we see every day and the advancement of knowledge. We have to understand the balance between hope and reality. Chloroquine is an old anti-malaria drug, along with its metabolite hydroxychloroquine. There is some evidence that this compound can reduce virus entry into cells, and this case suggests that it can be used as a prophylactic drug against infection and possibly in infected people.

Studies are ongoing, so there is no clear evidence that chloroquine can actually be effective. While waiting for scientific evidence, we have to remember that chloroquine, although quite safe, does have some important **side effects, especially regarding the heart**. It can lead to prolongation of the QT interval in the heart, leading to dangerous arrhythmias. This risk is higher in predisposed patients or in combination with other drugs. We know many DMD patients may have prolongation of QT intervals. It's fundamental to remember that chloroquine and chloroquine hydroxide have to be used under strict medical control and monitoring of heart function. Self-medication is not possible and highly risky.

Are there any drugs used when a subject is infected with COVID-19?

As anticipated, **there are no specific drugs available, since this coronavirus is a new virus**. A lot of progress has been made in the field of antiviral drugs for HIV infection, hepatitis, SARS and Ebola. Those viruses are somehow similar to the coronavirus as they are all RNA viruses. Even if they are different, it is hoped that some basic mechanism might be similar. This means that a drug developed against another virus might help in the case of this new coronavirus.

In case of COVID-19 infection, some drugs used for HIV or Ebola are being used off-label in infected patients. Off-label means that a drug is used for a disease outside its indication, but it can hopefully help, due to its mechanism of action, in patients for whom there is no specific therapy.

Following are a few examples of promising drugs. For each of them, anecdotal reports of beneficial effects in COVID-19 are available and clinical trials have been initiated and are needed to confirm efficacy. Remdesivir is a nucleoside analog developed with controversial results for Ebola infection.

Another treatment that raised interest is the combination of interferon alpha with inhibitors of viral proteases lopinavir/ritonavir. This is a cocktail used for HIV infection and can also be used in children older than two years.

Other antiviral drugs such as ribavirin with other inhibitors of reverse transcriptase (used again for HIV) have been considered with different levels of enthusiasm.

Another promising drug is favipiravir. This is an antiviral drug used for infection by the influenza virus and is available in Japan. It is being used off-label, and clinical trials are going to start in various countries.

Some antibacterial drugs can be considered in case of opportunistic bacterial infection in COVID-19. This is the case for azithromycin proposed in combination with chloroquine.

I would also mention tocilizumab, an inhibitor of interleukin-6 that has been approved for rheumatoid arthritis. It has great potential to control cytokine storms and has been used with success in Italy. Clinical trials are ongoing.

All these drugs are not for self-medication and are to be used under medical control. Many of them can only be used only in hospital settings. They can have serious side effects, especially in patients who already have a complex Standard of Care, such as DMD. So again, self-medication is very dangerous.

Other than searching for new specific drugs against COVID-19, intensive work is ongoing all over the world to create a vaccine, or to study plasma extract of patients to identify endogenous compounds that might be helpful. This is a very dynamic situation, things might change rapidly. We have to rely on robust scientific evidence.

Home4Duchenne & Home4Becker

Many pictures have arrived this week on **#Home4Duchenne** and **#Home4Becker**. It's important to continuously share pictures to raise awareness about staying home, especially for people with severe diseases like Duchenne. The impact of one person is important for the community. We received pictures of children from different places such as Spain and the United States, showing how they are living their quarantine. It's important that this campaign goes forward, so support your country and share your pictures with the hashtag.