



Updated Guidelines Regarding Coronavirus (COVID-19) to the FOP Community January 2022

The ICC is providing this update to the prior statement of March 2021. This document focuses on updated information on COVID-19 infection and updated guidelines on COVID-19 vaccination in FOP patients.

The Coronavirus (COVID-19) pandemic continues to pose significant risks to the population worldwide - including the FOP community - with new variants of SARS-CoV-2 virus emerging. **The ICC recommends that people living with FOP to continue to follow strict precautionary measures to prevent infection from SARS-CoV-2, the virus that causes the COVID-19 illness.**

The recommendations regarding COVID-19 infection and COVID-19 vaccinations to the general public are changing rapidly, are country and locale specific, and based on COVID-19 vaccine and treatment availability:

- **UNCHANGED!** The ICC does not provide recommendations on whether any individual patient with FOP should or should not receive a COVID vaccine. Vaccination for COVID should not be required for people living with FOP due to potential risk of flare up, but those who chose to do get the vaccination should follow the guidelines below. Discuss with your physician whether vaccination may be appropriate for you or your family.
- **UNCHANGED!** The decision to take a vaccine is a **personal one** and based on the balance of risks and benefits, and this should be discussed with your medical team. The ICC continues to recommend that COVID-19 vaccines be administered by the same route for which it was approved (i.e., intramuscular, if the vaccine was designed that way).
- **New!** Reviewing preliminary data shared by Dr Hsiao as a pre-print, [Social and Clinical Impact of COVID-19 on Patients with Fibrodysplasia Ossificans Progressiva - PubMed \(nih.gov\)](#)
 - o Among 15 FOP patients who received intramuscular COVID-19 vaccination, the most common symptoms were pain/soreness at the injection site, tiredness and

swelling which is similar to the reported symptoms in the general population. 1 out of 15 individuals with FOP that self-selected to receive the COVID-19 vaccination developed symptoms of a flare-up, which was managed by prednisone. None of the 15 patients required hospitalization.

- Among 10 FOP patients with COVID-19 infection, the most common symptoms were fatigue, loss of taste or smell and cough which is similar to the symptoms reported in the general population. 2 out of 10 FOP patients developed flare-ups and 1 FOP patient was hospitalized.
- Information regarding FOP and COVID-19 remains very limited and Dr Hsiao's paper does not include all FOP patients who have received the COVID-19 vaccines or had COVID-19 infection.
- **New!** In certain countries, vaccines are now available for children age 5 or over. The ICC does not provide recommendations on whether a patient with FOP should or should not receive a COVID vaccine. The COVID vaccines are not available in all countries for children aged 5 and over. Discuss with your physician whether the vaccine is appropriate for your child.
- **New!** ICC **does not provide recommendations for or against** the booster vaccination. Please consult with your medical team prior to receiving the booster to discuss if a booster is appropriate and safe for you.
- **New!** Patients with FOP are at high risk of complications with COVID-19 infection and should discuss with their medical team if the use of monoclonal antibodies or anti-retroviral medications would be beneficial, in the event of a SARS-CoV-2 infection.
 - Monoclonal antibodies are often given intravenously and are approved for adults and pediatric patients (specific populations and age ranges vary). Antibodies are in short supply and may require meeting specific criteria, including age, infection status, and risk of complications, as well as the specific strain of SARS-CoV-2. In general, patients with FOP are not considered immunocompromised, but are at increased risk from COVID due to pulmonary complications. These interventions should be started as early as possible and usually within 5-10 days of symptoms onset.
 - Anti-retroviral medicines (such as Molnupiravir or Paxlovid) are pills or injections that have been approved in some countries such as the United States and United Kingdom, and are used for decreasing the severity of infection. They may need to be administered within 5 days of symptoms onset. Paxlovid or other therapies may be available as pre-exposure prophylaxis for certain populations. Discuss with your primary care physician to assess eligibility and appropriateness.
 - Again, availability and recommendations of the use of these treatments are rapidly changing and country/location specific. Please consult with your local medical team for advice and access to these medications.
- **New!** Growing data indicates that patients who have had a prior COVID infection are not well protected from subsequent infections. Continuing universal precautions is important,

and patients should discuss with their physician if getting vaccinated/boosted is appropriate.

Important! Blood antibody testing is not considered a reliable measure of protection against SARS-CoV-2 infection. It measures that a patient has been exposed and had an immune response, but it is not clear how an antibody titer correlates with protection status.

Important! Please consider being part of the observational clinical study for patients who have been exposed or diagnosed with COVID, or who are considering the COVID vaccine. This study is led by Dr Edward Hsiao at UCSF. Please contact him at edward.hsiao@ucsf.edu if you are interested in learning more.

Important! If you decide to take the COVID vaccine, the ICC recommends:

DO NOT TAKE ANY VACCINATION IF YOU ARE HAVING A FLARE-UP; WAIT AT LEAST TWO WEEKS UNTIL AFTER THE FLARE-UP RESOLVES.

- **Discuss your plans with your doctor.** Review any potential allergies or prior reactions like anaphylaxis that you should consider before taking the vaccine. Based on the US CDC guidelines, a patient can receive COVID vaccination as soon as 2 weeks after a COVID infection.
- **Take the vaccine via the recommended route and dose** (ie **intramuscular** for the currently available vaccines). Safety and efficacy of taking an IM vaccine through the subcutaneous route is not known, and could cause a more unexpected inflammatory response or poor immune response, and is currently not recommended.
- If possible, take the vaccine in a **location that is already fused**, as the vaccines all appear to induce some local site reaction (arm pain and swelling). For example, if your left hip or right shoulder are fused, you should use the muscle around those sites.
- Patients with FOP should be **flare free for at least 2 weeks** prior to receiving the vaccine.
- Have the injection done by an experienced nurse, physician, or pharmacist.
- The clinician should be aware that patients with FOP may have hidden HO and thinned muscle at the site of the injection. Avoid injecting directly next to existing HO bone if possible.
- Prior to the vaccination, **have ibuprofen or acetaminophen available**. Also, have a course of prednisone for flares available.
- Prednisone and other glucocorticoids could decrease the efficacy of the vaccine, please consult with your medical team if you are on chronic prednisone or recently received a high dose of prednisone.
- Make sure your physician is familiar with the ICC Treatment guidelines, specifically on vaccinations and flare management. [Guidelines - International Clinical Council \(ICC\) on Fibrodysplasia Ossificans Progressiva \(FOP\) \(iccfop.org\)](#). Notify your physician you plan to do the vaccine, and when.
- On the day of the injection:
 - Your local team may not allow you to take ibuprofen or acetaminophen prior to the injection (this is because they may screen for COVID symptoms first).
 - After you receive your injection, there may be a brief observation period.

- After that is completed, take ibuprofen (2 to 3 times/day) or acetaminophen (2-3 times/day) following the label instructions, for the next 48 hrs, regardless of your symptoms.
- Consider local cold compress for 15 to 30 minutes after vaccination
- Rest and stay hydrated.
- In the event of a flare, contact your physician for guidance. You may need to do a short course of prednisone, but this needs to be balanced with the immunosuppressive effects of steroids. The usual flare dosing is prednisone 2 mg/kg/day up to 100 mg, for 4 days; your physician may recommend starting at a lower dose, depending on your symptoms.
- Even if you take the vaccine, you still need to continue physical distancing, wearing masks, and appropriate hand washing

The ICC can't guarantee that these steps will "work" to prevent complications. All medications and treatments have risk, so it is important to discuss your specific situation with your doctor as you decide whether to take the COVID-19 vaccine or not.

Make sure that you complete the full immunization regimen recommended (i.e., do both doses if the vaccine recommends 2 doses)

Discuss with your physician if you should have a booster and if that is appropriate for you.

COVID management is an area of active investigation and rapidly changing. The ICC will update recommendations as more information becomes available.

Important! How does the development of a vaccine change things?

- Recent developments of a vaccine for COVID-19 provides long term hope. However, the impact of the vaccine on the pandemic will take quite some time to manifest. Many types of vaccines are being tested around the world and new variants of SARS-CoV-2 virus are emerging and so this is a rapidly changing field.
- **Vaccines to date are not available for children younger than 5 years, or for anyone by subcutaneous delivery.** These are actively being studied, and will be added to the guidelines once available.
- The exact duration of immunity conferred by the vaccines is unknown but does not seem to be lifelong. Patients who had a vaccine over 6 months ago should consider a booster.
- At this time, the ICC does **not recommend vaccination for children younger than 5 years** due to the lack of safety and efficacy data.
- At this time, the **ICC does not provide recommendations for or against vaccination for patients with FOP older than 5 years of age.** The specific risks and benefits should be discussed with your physician.
- The ICC recommends that **FOP family members and caregivers get vaccinated for SARS-CoV2** if safely available for them.
- Vaccinations can take 2+ weeks to show efficacy, so there is no protection immediately after vaccination. In addition, vaccines do not confer absolute immunity to the SARS-CoV-2 virus, and may not have activity against all forms of the SARS-CoV-2 virus. **Anyone who receives a vaccine should still continue with masking, hand hygiene, and physical distancing.**

- This information is rapidly evolving. Please discuss with your local care providers regarding benefits and risk of any locally approved vaccines.
- It's very important to maintain social distancing and wearing a mask when around members outside your household
- Additional updates will be shared as new information becomes available.

Important! How do the new treatments for COVID infection change things?

- **There are several monoclonal antibodies and new small molecule drugs that are being used to treat patients with active COVID infections.** These are important for supplementing the vaccines.
- Vaccines work by decreasing the risks of infection, and decreasing the severity of the disease if infection occurs. The antibody and small molecule treatments help decrease the severity of COVID after infection occurs.
- Some of these medications may require intramuscular injections.
- **Some of the medications may be useful for preventing infection by SARS-CoV-2** (i.e., prophylaxis). However, to date these medications appear to be less effective than vaccination, have unknown safety data, and may have limited availability.
- **The antibody and small molecule treatments for COVID are in extremely limited supply.** They may not be available in your area or have strict limitations on who can receive the medication. It is important for each patient with FOP to discuss with their primary care physician about these treatments and to plan ahead.

Important! Recommendations if a patient with FOP or caregiver tests positive for SARS-CoV-2

- **Notify your primary care physician to help coordinate care**
- **Follow your local guidelines for isolation/quarantine and the needed durations and procedures.**
- Everyone, including the person with the positive SARS-CoV-2, should wear a mask at all times to avoid transmission.
- **Patients who are negative for SARS-CoV2 but have similar symptoms should be tested for influenza.**
- **Patients should use ibuprofen or acetaminophen, and other over the counter medications, to manage the upper respiratory symptoms.** Discuss with your physicians regarding the appropriate medications and potential medication interactions.
- **New!** Patients with FOP are at high risk of complications with COVID-19 infection and should discuss with their medical team if the use of monoclonal antibodies or anti-retroviral medications would be beneficial, in the event of a SARS-CoV2 infection or high risk exposure
 - o Monoclonal antibodies are given intravenously or intramuscularly and have various different approvals. Some of these antibodies are not effective against certain variants. Those interventions should be started as early as possible and within 5-10 days of symptoms onset. Contact your physician to discuss if these medications are available and beneficial.
 - o Anti-retroviral medicine (such as Molnupiravir) are pills or injections that have been approved in some countries, including the United States and the United Kingdom, and should be administered within 5 days of symptoms onset and positive COVID test. Right now, in the USA, Molnupiravir is only for adults older than 18 years of age.
 - o Again, availability and recommendations of the use of these treatments are rapidly changing and country specific. Please consult with your local medical team.