# UPDATED GUIDANCE ABOUT COVID-19 FOR PEOPLE WITH ATAXIA-TELANGIECTASIA AND THEIR FAMILIES

In consultation with:

**Howard Lederman, MD, PhD**
**Johns Hopkins Children's Center**
Baltimore, Maryland, USA
Director, Ataxia Telangiectasia Clinical Center
Director, Immunodeficiency Clinic;
Professor of Pediatrics and Medicine

## VACCINES

The COVID-19 vaccines are safe and effective and should be administered on the usual schedule for all people with A-T, their household members and other close contacts who are at least five years old (this age will decrease as the COVID vaccines are approved for younger children).

- The COVID-19 mRNA vaccines (Pfizer and Moderna in the U.S.) may be more likely to stimulate immunity in people with immunodeficiency and should be used in preference to the other types of COVID-19 vaccines (Johnson & Johnson in the U.S.).
- None of the COVID-19 vaccines contain live virus and thus cannot cause a COVID-19 infection.
- Data are limited, but we have seen no evidence yet that individuals with A-T are at an increased risk for any side effects from receiving the COVID-19 vaccines.
- The COVID-19 mRNA vaccines that are currently available in the US are about 90% effective in preventing hospitalization in the general population. They may not be as effective in a person with A-T, depending on the person's immune function.
- People with A-T should receive a COVID-19 vaccine whether or not they previously had a COVID-19 infection, and whether or not they are receiving gamma globulin therapy (such as IVIG or SCIG). The current lots of gamma globulin contain some antibody to COVID-19, but it is not known how much antibody is needed to provide protection.
- The COVID-19 vaccine is unlikely to provide protection to people with A-T who are undergoing chemotherapy or on a high dose of an immunosuppressing agent such as prednisone. People in this category should talk to their doctor about getting the vaccine, but it will likely not provide protection. Note, however, that A-T patients currently participating in the EryDel-sponsored clinical of EryDex (dexamethasone) for A-T should receive a COVID-19 vaccine, as the steroid dose given in that trial is probably too small to interfere with the vaccine.
- Even after receiving the vaccine, one cannot assume that it has generated anti-COVID-19 antibodies and protected a person with A-T from COVID-19 infection. Therefore, a blood test for levels of the anti-COVID-19 antibody should be performed four to six weeks after the last dose of vaccine has been received.
    - If the test for the anti-COVID-19 antibody shows a positive result and the person does not have a history of underlying lung disease, they may feel a little more comfortable about resuming their activities in the community, as long as they and the people around them continue to wear masks, and practice careful handwashing and social distancing. Antibody levels should be re-checked in four to six months to see if protective levels of antibody are maintained.
    - If the test for the anti-COVID-19 antibody shows a negative antibody result, the person with A-T should assume that they do not have protection and continue to maintain all current safety protocols.
- People with A-T should follow general community guidance for obtaining booster doses of vaccine, and whether all doses need to be from the same manufacturer.

For those in the U.S., Dr. Lederman has provided the following codes for tests to measure antibody response to COVID vaccines:

- **Quest 34499**
- **LabCorp 164055**

## CAN PEOPLE WITH A-T HAVE AN ALLERGIC REACTION TO THE VACCINE?

If anyone has a history of an allergic reaction to any drug, they should consult their doctor or allergist before receiving the COVID-19 vaccine. That being said, the rare, sudden allergic reaction to the vaccine (anaphylaxis) seen in some people who do not have A-T should not be a risk for A-T patients, because the majority of people with A-T do not have the IgE antibody that causes this type of reaction.

## DO WE NEED TO MAINTAIN SAFELY PROTOCOLS AFTER WE ARE VACCINATED?

We know that vaccines decrease the chance of severe infection but they do not prevent all infections. People who have received the vaccine can still spread COVID-19, even if they have no symptoms of infection. We do not know how long the vaccine protection will last. With that in mind, it is critical to continue wearing masks, carefully wash hands and maintain social distancing even after receiving the vaccine.

## DO A-T CARRIERS HAVE SPECIFIC RISKS WITH THE VACCINE

Biological parents and some siblings of people with A-T are A-T carriers and carry one copy of the mutated ATM gene. There is no increased risk above that of the general population for carriers regarding COVID-19 or the COVID-19 vaccines. For any specific concerns about contraindications (i.e., reasons not to receive the vaccine, such as allergies, autoimmune problems, etc.), check with your local doctor.

## WHAT HAVE WE LEARNED ABOUT COVID-19 IN PEOPLE WITH A-T?

So far, people with A-T don't seem to be at much higher risk for severe COVID than anyone else who is infected with COVID, but we have information about only a small number of people with A-T who have had COVID. To date, the A-T Clinical Center only knows of one person with A-T who had COVID and had a severe outcome (an acute deterioration in neurologic function).

## WHAT SIDE EFFECTS CAN BE EXPECTED?

The possible side effects of the vaccines (fatigue, headache, soreness at the injection site, low-grade fever or flu-like symptoms) should be no more likely or more severe in people with A-T or A-T carriers than in the general population.In general, vaccine side effects are less likely and less severe than the symptoms of a COVID infection in an unvaccinated person.

## SHOULD PEOPLE WITH A-T BE GOING TO SCHOOL?

Decisions about going to school have to be made on a case-by-case basis in consultation with your local health provider. The decisions need to take into account the rate of community spread, the steps that individual schools are taking to limit transmission (testing, policies about masks and vaccines, social distancing, class size) and the risk factors for severe disease in an individual person (immunodeficiency, lung disease, concurrent medicines, age, etc.)

## WHAT TO DO AFTER CATCHING COVID-19

There are currently three treatment options for outpatients who are infected with COVID-19 and are not severely ill. The decision about whether to use these drugs needs to be made on an individual basis, and the drugs should be used for people who have risk factors for severe disease if they are infected. Below are risk factors that apply to the general population, but also take into account the important features of A-T:

- Immunodeficiency (on gamma globulin therapy for antibody deficiency, on chemotherapy, on high-dose steroids or another immunosuppressive drug)
- Lung disease
- Obesity
- Cancer
- Over 20 years of age (this is very different than for the general population)

These treatments are authorized for use under the U.S. FDA EUA for people who are at least 12 years old and weigh at least 40 kg (about 88 lbs).

The available drugs include:

- **Sotrovimab** (GlaxoSmithKline) – This is a monoclonal antibody that is active against the currently circulating COVID-19 variants including omicron. It is administered by IV infusion that takes about 30 minutes. This antibody is in short supply and currently available only for people with COVID infection. It is not available for people who have been exposed and may or may not have been infected.
  - Note: The main side effect is an infusion reaction causing wheezing or other breathing problems, rash such as hives, and low blood pressure – all of which should be unlikely in a person with A-T. One other issue is that getting this infusion may prevent the person from developing their own antibodies to the COVID vaccine.

- **Paxlovid** (Pfizer) – This is a pill that should work against all of the COVID-19 variants. It requires taking as many as three pills twice a day for five days. There is a long list of drugs that may interact and should be avoided with Paxlovid, but few people with A-T are likely to be taking those drugs. The most common side effects are altered taste, diarrhea, high blood pressure and muscle aches.

- **Evusheld** (Astra Zeneca) – This is a long-acting combination of two monoclonal antibodies that is authorized for emergency use for pre-exposure prophylaxis (prevention) of COVID-19. It should be safe for people with A-T but is currently in extremely short supply in the U.S. It is given in 2 separate intramuscular injections.

- **Molnupiravir** (Merck) – This drug should not be given to a person with A-T, because of the chance that it may alter a person's DNA, and A-T patients have difficulty repairing certain kinds of DNA damage.

***Please note:*** *This guidance is based on what is known about available COVID-19 vaccines and treatments in the United States as of January 3, 2022.*

A-T CHILDREN'S PROJECT

Ataxia-telangiectasia (A-T) is a genetic disease that causes loss of muscle control and balance, cancer, lung disease and immune system problems in children and young adults, shortening their lives. The nonprofit A-T Children's Project partners with academic and industry investigators worldwide – organizing and supporting innovative research, conferences, clinical teams, data platforms and biomarkers – to optimize disease management strategies, develop new treatments and find a cure.

atcp.org - info@atcp.org