

ALD and COVID-19: Frequently Asked Questions

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What is COVID-19?

Coronavirus disease 2019, or "COVID-19," is an infection caused by a virus called SARS-CoV-2. COVID-19 was first identified in late 2019 in Wuhan, China. It has spread quickly since then to many parts of the world.

People with COVID-19 may develop flu-like symptoms such as fever, cough, and sore throat. Most people have mild or no symptoms and recover on their own. However, some people have severe symptoms, like difficulty breathing and pneumonia. Older adults and people with certain medical problems are at higher risk for having severe infection.

Experts are studying this virus and will continue to learn more about it over time.

My child was diagnosed with ALD through newborn screening. He is asymptomatic. Do we need to be concerned about him any more than our non-ALD children? Is he at any higher risk than a healthy, non-ALD child for COVID-19?

There is no evidence that an asymptomatic child with ALD, diagnosed by newborn screen or by family history, is at higher risk for COVID-19 related complications, but because this infection is new, we have limited data. It remains important to practice good hand hygiene, avoid touching the face and practice social distancing to limit the spread of COVID-19. We recommend looking at the CDC website for updates. Fortunately, most children who get COVID-19 do not appear to be at high risk for the severe forms of the disease. Healthy children with ALD probably have the same risk as healthy children without ALD.

It's likely that routine screening visits for adrenal insufficiency will need to be conducted remotely or delayed as physicians and other healthcare providers are asked to care for COVID-19 patients. It's important to know the signs and symptoms of adrenal insufficiency and to reach out to your pediatrician or pediatric endocrinologist if you notice changes in your child.

Signs of Chronic Adrenal Insufficiency	Signs of Adrenal Crisis
Darkening of the skin (Hyperpigmentation) particularly in skin creases, scars or gums	Severe fatigue
Fatigue, weakness	Nausea/Vomiting
Weight loss/Lack of weight gain/Failure to thrive	Abdominal pain
Decreased appetite/Anorexia	Dehydration
Intermittent abdominal pain	Low blood pressure/Fainting
Low blood pressure/Dizziness	Altered mental status
Loss of pubic hair (in adolescents/young adults)	Can be associated with other signs of infection
Rarely, if mineralocorticoid deficiency is present, salt-craving behaviors & increased urination	

If a child with ALD becomes infected by COVID-19, is he more likely to develop adrenal insufficiency?

Your child's adrenal function is especially important when he gets sick, because the adrenal gland helps to fight infection. Children with ALD can develop adrenal insufficiency early in life, even when they are only a few months old. The screening test for adrenal function is an ACTH level in the blood.

If your child becomes sick, have an ACTH level checked when they are assessed by their pediatrician. This is to make sure they have not developed adrenal insufficiency since their last ACTH test. This is different than what would happen in the child not affected by ALD.

If you already know your child with ALD has adrenal insufficiency, there are additional steps to take (see below).

Are adrenally insufficient patients considered high-risk?

There are reports that adrenal insufficiency is associated with a higher risk for infections, including respiratory infections. Much of the data related to higher risk for infections were collected in adults with autoimmune primary adrenal insufficiency. Adults with autoimmune adrenal insufficiency are also at higher risk for other medical conditions, including type 1 diabetes mellitus, which can also increase the susceptibility to more severe infections. The most important thing to do is prevent the infection; practice good hand hygiene, avoid touching the face and practice social distancing to limit the spread of COVID-19.

What should be done with steroids if the patient is exposed to COVID-19 and has adrenal insufficiency?

For exposure to COVID-19, there are no recommendations to change steroid doses. Routine daily doses should continue to prevent adrenal crisis. If a patient with adrenal insufficiency develops symptoms of COVID-19, they should follow the stress dosing recommendations that their prescribing provider has given them.

My son is adrenally insufficient. If he becomes infected by COVID-19, how do I figure out stress dosing for respiratory symptoms?

I recommend that you contact your son's primary endocrinologist to review sick day management recommendations. At this time, the recommendations for stress dosing for COVID-19 remain the same as with all other infectious processes. Increased doses of steroids should be given with fever and other significant complications.

My child is on steroids. Should I change his regimen?

If your child does not have COVID-19 and is prescribed steroids to treat adrenal insufficiency, the best course of action is to continue the current steroid doses. You should not change the doses of steroids without speaking to your child's prescribing provider. The daily doses of steroids are necessary to prevent an adrenal crisis.

What is a stress dose of steroids, and when should I do it?

Stress doses of steroids are intended to mimic the normal increase in cortisol (the body's "stress hormone") that the adrenal glands should make when the body experiences physical stress.

Physical stress occurs when a patient is sick, has fever, breaks a bone, has a seizure or requires surgery/anesthesia. Ideally, stress dosing instructions should be reviewed with the primary endocrinologist prescribing the steroids.

How should I prepare for COVID-19 and other emergencies?

Due to concerns for significant shutdowns, we recommend all patients with adrenal insufficiency and their families make sure they have and review the following:

- Have at least one month of steroid medication (including enough for stress dosing) at home.
- Make sure you have an emergency Solu-Cortef Act-O-Vial, as well as needles and syringes for administration. Check the expiration dates.
- If you don't have a month of medication or Act-O-Vial, or if your refills will expire in the next month, call your prescribing provider's office to request refills.
- Review sick day management plans.
 - Make sure you have the emergency contact information for your endocrinologist
 - If you don't have written plans, ask your primary endocrinologist's office to send you a copy of the written plan. This will be important to review with on-call and emergency room providers to guide treatment with stress doses of steroids.
 - I recommend patients/parents take pictures of written plans with their phone and enter emergency contact information into the phone (it's easier to find a phone than papers when you are in a rush).
 - Specifically for COVID-19, practice social distancing, wash your hands and avoid touching the face. Follow CDC and your state Department of Health recommendations.

Does COVID-19 increase the risk of cerebral ALD?

Good news, we don't believe that COVID-19 would put your child at any higher risk of developing cerebral ALD. Remember that COVID-19 is a new infection, so we don't have a lot of direct experience yet. But, COVID-19 is a respiratory virus, and we know that other more common viruses (like those that cause colds or the flu) **do not** increase the risk of cerebral ALD. We are not recommending any change in the MRI screening schedule.

Are ALD patients who have had a bone marrow transplant (BMT) or gene therapy considered high-risk if they get COVID-19?

Yes. Children who have recently undergone BMT or gene therapy within the last 2 years have weakened immune systems, which puts them at risk for more severe infection. Children who have graft-versus-host disease (GVHD) can also have a flare up of their GVHD with an infection and are at higher risk for infection.

Should BMT or gene therapy be delayed for patients with cerebral ALD during the COVID-19 pandemic?

Since early BMT is the safest and most effective therapy for cerebral ALD, transplant should not be significantly delayed when indicated. However, hospitals are taking extra precautions during the COVID-19 pandemic to prevent the spread of infection to patients who need to be in the hospital.

Our son has a tracheostomy. Are there differences in tracheostomy care? How should we guard against risks for infection?

Pulmonologists recommend following all of the general guidelines for COVID-19. This means strict hand washing and no trach care by anyone with cough, cold, or fever.

My son is fully dependent and bed ridden. What would we need to tell the doctor if he became sick with COVID-19?

The main thing you should let the doctor know is that patients with ALD can have adrenal insufficiency that requires hormone replacement to help fight infection. He may also be at higher risk for severe infection because of low lung function.

Are men with AMN considered high-risk if they get COVID-19?

It is difficult to generalize for men with AMN, because everyone is different. But some AMN patients may be at higher risk for severe infection due to their chronic neurological symptoms, poor mobility (i.e. wheelchair bound), and adrenal insufficiency.

Does being in a wheelchair make men high-risk?

Being wheelchair bound brings risks regardless of COVID-19, such as higher chances of urinary tract infections, ulcers, and falls when moving in and out of the wheelchair. It is especially important to wash your hands after using a wheelchair.

Are women with ALD considered “immune-suppressed” and do they get the virus easier?

We do not think that women with ALD are more susceptible to getting the COVID-19 virus. Women with ALD generally **do not** have adrenal insufficiency. Hence, their immune system is just as strong as that of people who do not have ALD.

Are individuals with ALD at higher risk of complications from COVID-19?

ALD and AMN do not affect the heart or lungs, so ALD patients with normal adrenal function should not have more trouble than others in fighting infections such as COVID-19. However, we are still learning about the clinical course of the virus, and there are many things we do not know.

Should individuals with ALD wear a mask outside the house?

The CDC does not currently recommend masks for use outside of specific medical settings. COVID-19 appears to be spread primarily through respiratory secretions during prolonged (>30 minutes) close contact (<6 feet) of an infected person. There is also a huge shortage of masks for all persons, including hospital staff caring for COVID-19 patients in the U.S. and worldwide. Following the CDC guidance, the best ways to protect yourself are hand washing and social distancing (maintaining 6 feet or more between you and all persons who are not part of your household). This means that you should avoid play dates, public playgrounds, and parks.

Do any recommendations change for patients who have ALD?

Make sure your adrenal function has been checked and any insufficiency has been adequately addressed by replacement therapy.

Should we cancel important appointments such as MRIs? What precautions should we take when we go to the clinic or hospital? How do I keep my child safe?

If you have to go to a hospital, talk to your provider about whether routine tests can be delayed or performed locally. Also discuss with your care team to see if any of these appointments can be done virtually. If your child needs regular bloodwork, talk to your provider to see if this can be done locally.

We have home nursing (care, infusions). Should I allow the nurse to enter the home? What precautions should we take?

Nurses should wear masks to limit the chances of passing on or contracting the virus. You should try to limit the number of nurses and other caregivers coming in and out of the house.

My child has weekly appointments at the hospital. Should we cancel or keep going?

After bone marrow transplantation, individual risks and benefit should be considered. Please discuss this with your provider.

Should I send siblings or spouses to school or work? Should I quarantine my spouse who works outside the home from my child?

Generally, it is advisable to limit contact, particularly if there is a patient in the house who is at higher risk due to neurological or endocrine symptoms. If you have a child that is bedridden, consider having the parent stay at home and be the primary caregiver. We understand that this is not always possible.