

**AHA COVID-19 Clinical Guidance Series**  
**Adult Congenital Heart Disease**

**KEY ON-AIR CONTRIBUTORS:**

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**Speaker 1:**

Welcome and thank you for joining us for this podcast brought to you by the American Heart Association. This podcast is part of a series focused on sharing information with healthcare providers who are caring for patients during the COVID-19 pandemic.

**Speaker 2:**

Hi, this is your power bite for the ACHD and COVID-19 infection podcasts of the American Heart Association with Eric Krieger and Anitha John. Today we're going to be discussing COVID-19 infection in the ACHD population. We'll talk about which populations we think are at greatest risk for bad outcomes from a COVID-19 infection, treatment considerations for medical therapy for COVID-19 in the ACHD population. We'll discuss how to manage outpatients with congenital heart disease in a positive COVID-19 test as well as considerations for inpatient management for ACHD patients with COVID-19. We'll give you an update on ongoing research projects and point you in the direction of resources that you can use to learn more about adult congenital heart disease and put you in contact with an adult congenital heart disease provider who may be able to provide advice for you if you're caring for hospitalized ACHD patients with coronavirus.

**Dr. Anitha John:**

Welcome to the Adult Congenital Heart Disease and COVID-19 podcast. I'm Dr. Anitha John. I'm the Director of the Washington Adult Congenital Heart Program at Children's National Hospital in Washington, DC. I'm here today with Dr. Eric Krieger, Director of the Seattle Adult Congenital Heart Disease Program, a joint program between University of Washington and Seattle Children's Hospital. Thank you to the American Heart Association for inviting us to discuss how the COVID-19 pandemic impacts patients with adult congenital heart disease.

**Dr. Anitha John:**

We're hoping today to address what steps providers can take when testing, treating, and following ACHD patients with a suspected or diagnosed COVID-19 infection. Adults with congenital heart disease are a growing population, as many folks know. It is now estimated that there are over 1.5 million adults with CHD alive in the United States. There are many different types of congenital abnormalities with lots

of different types of longterm complications, but in general, most patients do have higher risk of comorbidities such as arrhythmias, heart failure in addition to non-cardiac comorbidities such as kidney and liver dysfunction.

Dr. Anitha John:

We're going to be talking today about some specific ACHD conditions that might respond differently to Coronavirus infection or might be more susceptible to a poor clinical outcome as a result of infection. In addition, we're also going to discuss some treatment considerations for ACHD patients with COVID infection and also talk about some resources that providers and also patients can access for guidance. So let's start off the discussion with, I think, a really important question and one that's come up from a lot of providers and also patients as well too. How are ACHD patients different from other cardiac patients and what are some of the unique aspects that healthcare providers should be aware of?

Dr. Eric Krieger:

Thanks, Anitha. So as we know there's a wide variety of different adult congenital heart disease diagnoses out there and there's a number of different ways you can categorize them either by their anatomic complexity or the patient's physiologic state, meaning their residual hemodynamic sequela and their clinical state. I think right off the bat we need to acknowledge that we don't actually know how COVID-19 infection is going to affect different adult congenital heart disease subpopulations because it's a new pandemic and this question hasn't yet been studied, but we can try to take what we know about COVID-19 infections and what we know about different patients with congenital heart disease and speculate about some populations that may be particularly susceptible to bad clinical outcomes.

Dr. Eric Krieger:

So congenital heart disease patients in general are younger than the general population and that overall is favorable for ACHD patients because we know that COVID-19 infections tend to be more severe in older patients. That being said, ACHD patients have a number of comorbidities that have been associated with adverse outcomes from a COVID infection. For example, we know that hypertension was identified early in the Wuhan cohorts as a major risk factor for poor outcome from COVID infections and certain ACHD populations such as those with coarctation of the aorta we know are predisposed to hypertension, so we think that ACHD patients with hypertension as an underlying diagnosis, particularly coarctation may be susceptible to bad outcomes.

Dr. Eric Krieger:

A portion of ACHD patients related to their initial diagnosis or perhaps late repair have pulmonary arterial hypertension as a complication of their congenital heart disease and we can speculate that this is going to put patients at higher risk for bad outcomes because they have considerably less pulmonary reserve than patients without pulmonary hypertension. This would be even more pronounced, for example, in a patient with Eisenmenger's syndrome with baseline cyanosis. In that patient, a severe lung infection could certainly bring them into a critical. Another population that's worth mentioning is those who have had a Fontan operation for single ventricle anatomy.

Dr. Eric Krieger:

These patients have no ventricle pumping through the pulmonary circulation and are really vulnerable anytime they have a lung injury, lung infection, and pneumonia because they don't have the cardiac reserve to drive blood through the pulmonary circulation. But it's important to know that it's not just

these complex physiologies, even patients with seemingly straightforward anatomy like tetralogy of fallot. A lot of these patients are going to have restrictive lung disease from sternotomies and multiple thoracotomies, that may put them at increased risk of adverse outcomes. We know that the adult congenital heart disease population has a high prevalence of sleep apnea. Up to a third of ACHD patients may have sleep apnea, and recent studies have shown that sleep apnea does confer a much higher risk of adverse COVID infection. And we're hearing anecdotally, at least from our colleagues in New York, that many of the young patients who are hospitalized or intubated do have sleep apnea or obesity as an underlying condition. So those are just some of the conditions that I think are highlighted that may put patients at risk for an adverse outcome.

Dr. Eric Krieger:

One other group I do want to mention is those ACHD patients who have underlying heart failure or maybe those that are predisposed to heart failure. We're seeing that maybe 10 to 20% of patients with a COVID infection are having an elevated troponin level, and those patients seem to have a much worse outcome. It's not clear yet whether this troponin elevation reflects direct myocardial injury or myocarditis from the Coronavirus or whether this is just troponin release in the setting of sepsis or a cytokine storm or critical illness, but in either case, it's possible to speculate that patients with underlying ventricular dysfunction or, for example, patients with the systemic right ventricle is a consequence of transposition of the great arteries may be more susceptible to myocardial injury or myocardial damage associated with Coronavirus infection. So Anitha, I'll turn the next question back to you. Which patients do you think ought to be tested for Coronavirus if they're calling in describing symptoms?

Dr. Anitha John:

Thanks, Eric. That's a common question that many of our patients are having in addition to many of the primary care physicians who are taking care of them as well. If you look at the CDC website, it's [www.cdc.gov](http://www.cdc.gov), and look at the testing indications, you can see that the CDC advises that not all patients with suspected COVID require testing. Patients with mild disease and those who are clinically well can usually be treated with self-quarantine. That being said, there are a set of testing priorities, so obviously patients who are hospitalized and healthcare workers with symptoms should obviously be tested. Most of our patients I think fall into the next priority level, which is testing in patients who are at high risk of complications. These include older patients and those with longterm comorbidities. Heart disease is listed as a longterm comorbidity that can increase the risk for serious Coronavirus infection and I think that's where most of our ACHD patients would end up falling. Although as you've stated, we don't have a really a tremendous amount of data looking at congenital heart disease patients with COVID-19 infection.

Dr. Anitha John:

The next priority level includes those patients with underlying conditions and mild symptoms and so many of our patients would probably also fall within that priority level as well. It's really too early to know which patients are at highest risk, but certainly those patients who have many of the features that you just mentioned in addition to any other underlying comorbidities such as hypertension, heart failure, arrhythmias should really be considered for testing. Some patients have alerts that have been created by their various centers that identify which patients should be tested, but really for the most part, I think it needs to be a clinical judgment in terms of which patient should be referred. The reality is, is that testing is really determined by local availability.

Dr. Anitha John:

Some places have widely available rapid testing, other places do not. There are some states that allow for testing without a doctor's order and other states that do not. Again, I think clinical judgment really needs to be utilized as to what type of testing modality is used. Another consideration is really thinking about what the patient's underlying risk is and what risks might be conveyed by the actual testing itself. So there are some testing sites that might confer increased risk of exposure, and if you have patients with some of the features that you just mentioned, it might be worthwhile to really try and have them tested at a place that has a better turnaround time and less exposure, such as a drive through. Many hospitals have faster turnaround time, but that again varies depending on the center and the region.

Dr. Anitha John:

So Eric, let me turn back to you again. What do you do for COVID-positive ACHD patient who is at home, may or may not have mild symptoms? Where do you make the decision on when to bring them into the hospital?

Dr. Eric Krieger:

So the good news is that the majority of patients who test positive for COVID-19 infections are able to stay at home. And it's only a minority of whom require hospitalization, so that's good news. And it's been our limited experience, at least that this also holds true for ACHD patients with COVID-19 infection. The majority of the ACHD patients around the country who have tested positive have been able to recover at home and it's been uncommon for them to require hospitalization. So what that means is that most ACHD COVID positive patients will be at home. I don't think there's a specific subset of ACHD patients that I would recommend coming into the hospital just because of their underlying diagnosis. I might be more inclined to do that with an Eisenmenger patient, I suppose, but I still think we need to make the decision about whether to hospitalize a patient based on the course of their infection, and patients with mild symptoms don't need to be hospitalized and patients with more severe symptoms do need to be hospitalized.

Dr. Eric Krieger:

That being said, if the ACHD patient is being followed at home, we think that they need to be followed quite carefully. There's been a lot of reports of patients who seem to be recovering reasonably well, but then clinically deteriorate midway through their COVID infection and that highlights the need for us to remain in regular contact with ACHD patients who test positive for COVID. And to me what that means is when feasible, that means regular telephone contact from the ACHD provider to the patient, and what we do here at the University of Washington is, we're calling and we're using a standardized questionnaire to ask about symptoms such as shortness of breath, chest pain, cough and fever. And we're asking those patients to rate their symptoms on a scale of one to nine and then documenting their results in the electronic medical record so that we can trend the patient's clinical course and potentially identify worrisome trends that might prompt us to hospitalize the patients.

Dr. Eric Krieger:

Patients that have more than mild symptoms or those with complex disease like pulmonary hypertension or Fontan circulation may require daily telephone check-ins. And these can be brief, but I think, as is often true in medicine, catching issues before they become problematic and before a patient becomes critically ill is important. Anitha, when we do hospitalize a patient, what do you think that the

ACHD team can provide in terms of consultation expertise? What do you think the in-patient team needs to be aware of specifically for the hospitalized ACHD patient with a COVID infection?

Dr. Anitha John:

Well, Eric, I think for patients who are hospitalized, it's really important to have an early initial consultation with an adult congenital heart disease specialist. And this is really important to define what the underlying anatomy is and to discuss some potential treatments and what might be some potential mechanisms of decompensation that should be watched for. If there are places that do not have an adult congenital heart disease specialist, folks can find the closest clinic by visiting the Adult Congenital Heart Disease website. That's [www.achaheart.org](http://www.achaheart.org). There is a very comprehensive clinic directory that is listed that has most of the clinics within North America. Any of those centers would be able to offer additional input and expertise if folks who are looking for help. And the reason for this really is, thinking about our patients, the fact that they have such very different types of anatomy, there can be specific concerns that may not be obvious to folks who don't take care of these patients all the time.

Dr. Anitha John:

So for example, patients with repaired aortic coarctation or those a prior classic Blalock-Taussig shunt, those are patients that may have inaccurate blood pressure measurements in one of their arms and this can lead to incorrect assumption that a patient is hypotensive. So really accurately identifying where a blood pressure should be measured or where an arterial line should be placed is really quite critical for management, especially if you have a patient who is critically ill and an adult congenital heart disease specialist really would be able to help troubleshoot some of those considerations. For very ill patients with COVID, I think ECMO still remains a consideration and given the fact that there are a variety of different anatomical considerations, ECMO might be very difficult, especially for folks who are not akin to really taking care of these patients and understanding what their anatomy is. So early consultation with an adult congenital heart disease specialist is needed.

Dr. Anitha John:

Sometimes consultation even with a congenital cardiac surgeon is also needed as well. And then you know really considering transferring the patient to an adult congenital heart disease center, especially for those with complex congenital heart disease and multiple comorbidities may be really the best care for the patient, but at the very least having some sort of phone consultation with the patient's adult congenital heart disease provider can really help identify some of these potentially issues.

Dr. Anitha John:

So Eric, understanding that there aren't really any definitive therapies for COVID-19, what are some of the potential considerations that people should really think about in adults with congenital heart disease?

Dr. Eric Krieger:

I think that we're still not sure about what therapies are effective in treating COVID infection overall, whether the antivirals are going to work or hydroxychloroquine or antibiotics or treatment with convalescent plasma. I think, in general, treatment should be guided by the treatment strategies, clinical trials and treatment algorithms used at an individual hospital and those should be applied to ACHD patients just like they're applied to patients without congenital heart disease. The only specific consideration that I can think of as it relates to direct therapy for COVID infection is just remembering

that hydroxychloroquine can be a QTc prolonging drug and that ACHD patients are prolonged to arrhythmia and many of them have an underlying bundle branch block on their ECG, so we just need to account for that and correct for that as we're following QT intervals with hydroxychloroquine treatment. I don't think I would withhold any of these medications for someone with ACHD or for someone with an underlying bundle branch block. It's just consideration that you need to take into account.

Dr. Eric Krieger:

Another thing that we're really starting to appreciate more and more over the last week or two is the very high rate of thromboembolic complications in hospitalized patients with COVID infection. That I think this is becoming a major source of morbidity and mortality in hospitalized patients in the United States. And centers that have high inpatient COVID censuses are starting to use therapeutic anticoagulation for large subsets of their patients, doing regular D-dimer testing on patients, and having a low threshold to therapeutically anticoagulate patients with COVID infection. It's important to remember that ACHD patients, some of them are predisposed to thromboembolic complications at baseline. For example, patients with Fontan and sluggish flow may have increased thromboembolic risk, patients with prosthetic heart valve. So we should just have a low threshold for initiation of therapeutic anticoagulation for our patients who we think are at higher than average risk of thromboembolic complications. But in general, I think we should be following the same treatment guidelines and treatment algorithms that are being used for patients without congenital heart disease.

Dr. Eric Krieger:

So just switching gears a little bit and transitioning into a different issue, Anitha, can you comment sort of onto the nonmedical impact of COVID-19 on our ACHD patients? A lot of these patients have experienced stress and trauma from having multiple heart surgeries and a lifelong medical condition. Do you have any experience or insight as to how this pandemic is impacting our ACHD patients from the psychological perspective?

Dr. Anitha John:

Thanks, Eric. I think that's a really important question. Adult patients with congenital heart disease have a high rate of underlying mood disorders. This includes anxiety, depression, PTSD. I know from our experience here, many of our patients really have had a lot of sort of reactivation type symptoms and for many patients, this COVID-19 pandemic has been very triggering and anxiety provoking, so it's really important to remember that some patients may need some additional resources during this difficult time.

Dr. Anitha John:

For our program, we've been referring our patients to our program social worker and have made additional referrals to both therapists and mental health professionals as needed. Some patients may need additional medical therapy during this time as well too, so that is also something to consider. A lot of patients are looking for additional resources on how to sort of help manage stress and anxiety. Some additional resources can be found on the CDC website at [www.cdc.gov](http://www.cdc.gov) on the Coronavirus page. In addition, the Adult Congenital Heart Association also recently held a webinar directed towards patients with regard to stress and anxiety management during this difficult time, and this can be accessed really to anybody. It can also be reached at [www.achaheart.org](http://www.achaheart.org) on the COVID-19 resources page. There are a lot of different resources that are on that page for both patients and providers, including several webinars and Q and A sessions. One additional resource of commonly asked questions in addition to

CHD-specific recommendations and resources was created by the various professional inpatient congenital heart disease group on the Congenital Heart Public Health Consortium website and that can be found at the [CHPHC.org](http://CHPHC.org).

Dr. Anitha John:

So as to kind of wrap up, what particular actions or key takeaways should we really take away from all of this, Eric?

Dr. Eric Krieger:

Yeah. Well one thing is that as we said at the beginning, we're speculating a little bit about how COVID infection is going to impact the ACHD population and it's certainly an area that needs research. There is currently a large multicenter registry trying to collect information on the outcomes of ACHD patients who are infected by COVID-19. It's being led by Jamil Aboulhosn at UCLA. Craig Broberg at OHSU is deeply involved. So far there's 21 centers that have IRB approval and are actively adding patients and another 69 centers worldwide that are in the process of obtaining approval. So that's great news and we should hopefully have some more information soon about this.

Dr. Eric Krieger:

So as we wrap up, just some key things to remember, that congenital heart disease patients have a lot of variability in their disease burden and their comorbidities, particularly for patients with complex forms of disease. It's important to involve ACHD specialists early. There's a resource out there that lists all the congenital heart disease centers around the country and phone contact information, if you do need to reach someone. Again, that's [www.achaheart.org](http://www.achaheart.org) and that will help put you in touch with an ACHD provider if you do have questions. So thank you very much and it's been a pleasure to talk with you today and wish everyone luck for the remainder of the COVID pandemic.

Speaker 1:

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