

**ARTICLE**

Managing the Adult Congenital Heart Disease Patient in the COVID-19 Pandemic—A New York Perspective

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ABSTRACT

Adults with congenital heart disease (ACHD) are likely at increased risk for complications of COVID-19. ACHD centers should prepare to deliver routine cardiac care and support for patients with COVID-19 safely at home, as the number of COVID-19 infections worldwide continues to increase. This brief report aims to share the strategies we have used in our ACHD program to manage and treat our patients during this global health crisis at one of the initial epicenters of the pandemic in New York City, and offer suggestions for preparation for ACHD clinicians.

KEYWORDS

COVID-19; SARS-CoV-2; coronavirus; adult congenital heart disease; infection

1 Introduction

The severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2 or COVID-19) pandemic has presented unprecedented challenges for healthcare professionals internationally. Patients with pre-existing cardiovascular disease are at increased risk for worse outcomes when infected with COVID-19 [1]. Adults with congenital heart disease (ACHD) represent a growing population of primarily young adults whose comorbidities likely put them at an inherently high risk of complications from the COVID-19 infection [2]. This brief report delineates the strategies we have used in our ACHD program to manage ACHD patients during this global health crisis at one of the initial epicenters of the pandemic in New York City.

2 Preparing the ACHD Medical Team

2.1 High Risk Patient List

As ACHD centers prepare for local fluctuations of COVID-19 cases over the coming months and varying degrees of stay-at-home orders, there are actions the medical team can take to ensure safety and anticipate patient needs for both routine cardiac care and care for patients with COVID-19 (Tab. 1). Creating a list of patients at increased risk for severe outcomes of COVID-19 will allow ACHD clinicians to proactively monitor these patients with virtual visits or check in phone calls. Patients with complex congenital heart disease (Anatomy Stage III or Physiological Stages B, C, or D according to ACHD Anatomy and Physiological Stage Classification) are considered to be at increased risk for complications



of COVID-19 [2,3]. Additional patients who may be considered high risk include (a) patients with respiratory diseases such as restrictive lung disease, asthma, hypoplastic lungs, cyanosis, or recurrent pneumonias, (b) patients who are pregnant, and (c) patients who need procedures or surgeries in the near future. Furthermore, there is concern that patients are hesitant to report cardiac symptoms, follow routine cardiac care, or go to the hospital for cardiac emergencies due to fear during the pandemic. The ACHD medical team should encourage patients to seek prompt medical attention as needed to avoid cardiovascular complications.

Table 1: Adult Congenital Heart Disease Program COVID-19 Preparedness

<p>Preparing the ACHD Team</p> <ol style="list-style-type: none"> 1. Create a list of high-risk patients and schedule televisits or phone calls for early symptom detection and continuity of cardiac care 2. Establish telemedicine workflows and ensure patients have home monitoring devices, including blood pressure cuffs, pulse oximeters and scales 3. Review list of patients on warfarin to determine who can participate in INR self-checks, delay INR checks, or transition to NOACs 4. Establish plan for home cardiac event monitors with QTc capabilities and home lab-work 5. Coordinate in-person visits and testing to be at the same day and location to reduce risk of exposure 6. Ensure adequate PPE and COVID-19 screening for staff
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<p>Preparing the ACHD Patient</p> <ol style="list-style-type: none"> 1. Inform patients of current recommendations for safety and establish an action plan to follow if patient or family member develops symptoms 2. Offer 90-day prescriptions for medications or transition to delivery pharmacies 3. Reinforce the use of medical passports for patients in the event of hospitalization 4. Provide resources for mental health support at home
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<p>Action Plan for the Suspected COVID-19 Infected ACHD Patient</p> <ol style="list-style-type: none"> 1. Administer diagnostic testing as soon as possible when symptoms are suspected to aid in early detection and management 2. Low threshold for acquiring blood cultures to rule out bacterial endocarditis 3. Conduct frequent check in calls for patients with COVID-19 4. Utilize home pulse oximeters and scales to monitor for hypoxia and titrate diuretics, respectively 5. Reinforce medication compliance of routine medications, particularly anticoagulation

NOACs = non-vitamin K antagonist oral anticoagulants; INR = international normalized ratio; PPE = personal protective equipment.

2.2 Access to Care

Developing workflows and innovative strategies to ensure patients have access to care is a crucial component of preparedness for the medical team. Telemedicine is an effective intervention to provide care for cardiac patients safely at home [4]. It is important that ACHD teams have adequate technology and access to virtual information technology (IT) support. Patients participating in telemedicine should be encouraged to obtain pulse-oximetry devices, blood pressures cuffs and scales, depending on their condition, in order to undergo routine cardiac monitoring at home.

Additional opportunities to provide continuous cardiac care at home include at-home lab draws and cardiac event monitors that can be mailed to the patient. It may be helpful to have cardiac event monitors with QTc monitoring capabilities as some medications experimentally used with COVID-19, such as lopinavir/ritonavir prolong the QTc interval [5,6]. Previous QTc prolonging agents such as hydroxychloroquine and azithromycin are no longer recommended at this time [7–9]. Other commercially

available rhythm monitoring devices and watches may be useful, although limited data is available at this time. ACHD centers that provide testing in the hospital can consider offering testing at satellite or local locations to reduce risk of exposure.

Urgent care centers have increased in popularity as an alternative care site for non-emergent conditions [10]. The COVID-19 pandemic has also led to an increase in utilization of technology for early evaluation of respiratory symptoms [11]. ACHD centers should develop a relationship with urgent care centers and virtual urgent care to evaluate their patients and collaborate to make decisions about testing, care escalation, and treatment. As it may be challenging in the ACHD patient to distinguish between COVID-19 and cardiac symptomatology, the current pandemic should raise high level of suspicion of COVID-19 complicating a baseline heart condition.

The use of patient-centered medical homes and care coordination, which have always been mainstays in care of the ACHD patient, is crucial during the COVID-19 pandemic in order to reduce risk of exposure [12,13]. For patients that require in person visits or testing, ACHD teams should coordinate care for visits with multiple providers or multiple testing modalities on the same day at the same location to minimize risk of exposure.

2.3 Anticoagulation

Clinicians must address patients requiring anticoagulation in a timely fashion to ensure they can safely remain anticoagulated. Anticoagulation is recommended for many patients with congenital heart disease for a variety of reasons including mechanical heart valves, arrhythmia, and prior thrombotic events, amongst others [3]. ACHD centers can collaborate with anticoagulation clinics to offer self-testing for INR at home, as telemedicine and oral anticoagulation management with self-testing are suitable and safe options [14]. Patients with stable INR can be offered extended INR testing with an interval of up to 12 weeks [15]. The ACHD guidelines recommend anticoagulation with vitamin K antagonists (VKA) in adults with Fontan palliation with known or suspected thrombus, thromboembolic events, or prior atrial arrhythmia, and no contraindications to anticoagulation [3]. Given the challenges of VKA monitoring during the pandemic, and based on a recent large international multi-center prospective study demonstrating safety and efficacy of non-vitamin K antagonist oral anticoagulants (NOACs) in Fontan patients, NOACs may be considered instead of VKAs on a case by case basis [16].

3 Preparing the ACHD Patient

An important aspect of ACHD care during the COVID-19 pandemic is educating and empowering patients with the information they need to reduce their risk of infection and maintain their health at home. ACHD clinicians should keep their patients informed with current guidelines and recommendations for staying safe during the COVID-19 pandemic, in addition to what to do if a family member or roommate develops viral symptoms. Patients can be offered 90-day prescriptions for medications or transition to delivery pharmacies to reduce their risk of exposure by traveling frequently to the pharmacy.

Outreach and educational programs are important components of care for ACHD patients, as some patients exhibit a lack of awareness about their condition in general [3,17]. Because of the novel and evolving nature of the COVID-19 pandemic, patients may be unaware of symptoms and less likely to report symptoms to their provider. For example, patients may be unaware that gastrointestinal symptoms appear to be more common in presentation in New York City than in China during the initial outbreak [18]. Patients should also be educated about loss of sense of taste and smell as a common symptom associated with COVID-19 [19]. Additionally, empowering patients with an action plan for what to do if they develop symptoms may increase the likelihood of effective and efficient care, as well as ease anxiety. This action plan must include alerting the ACHD team, as involving ACHD specialists in patient care improves outcomes [3]. Patients should keep in their wallet or on their mobile phone relevant health information easily accessible for emergency situations to be provided to emergency personnel or clinicians at an outside hospital, including their past medical history, medications, and ACHD team contact information, which patients may already have in their medical passports [17].

As many patients are confined to their homes, they are often exercising less, which leads to deconditioning. Patients may report an increase in palpitations and lightheadedness, as those who are deconditioned are more likely to experience orthostatic hypotension and sinus tachycardia. ACHD clinicians should encourage patients to stay active at home and set daily step goals. Additionally, ACHD patients have an increased prevalence of anxiety and depression, which is associated with worse cardiovascular outcomes [20]. The COVID-19 pandemic, similar to other public health emergencies, has increased the need for psychological support for individuals across the country [21]. ACHD clinicians can collaborate with social work to prepare a toolkit of resources for mental health support at home. Telemedicine may also play a role in managing psychological health for patients at home [22].

4 Medical Care of the ACHD Patient with COVID-19

As the number of patients with COVID-19 and ACHD inevitably continues to grow, we aim to share the strategies we have used to care for our increasing number of infected patients. Patients should undergo diagnostic testing as soon as possible when symptoms are suspected to aid in early detection and management. Because testing may be limited or untimely in certain regions of the country, diagnosis of COVID-19 may be based on clinical evaluation, close contact and prior travel history, regardless of testing [18]. Although COVID-19 may be the top differential diagnosis based on clinical presentation and current local transmission rates, it is important to consider a wide differential, including bacterial endocarditis. Patients with the highest risk for endocarditis include those with previous history of infective endocarditis, prosthetic heart valve replacement, cyanotic congenital heart disease, and cardiac implanted electronic device [23]. Endocarditis and COVID-19 have some similarities in their clinical presentation, such as fever, fatigue and myalgia. Therefore, the differential diagnosis of subacute bacterial endocarditis should be taken into consideration with a low threshold for taking blood cultures in the febrile patient.

ACHD programs should offer daily to weekly check in phone calls to patients who have COVID-19, depending on severity of viral symptoms and congenital heart disease, to closely monitor the patient and provide recommendations. In an effort to identify cases of COVID-19 in ACHD patients at the earliest stage, check in phone calls may also be made to patients with roommates or family members that are suspected of having COVID-19. Home daily oxygen saturation monitoring, especially in patients with baseline desaturation, is valuable for diagnosis of COVID-19 related to pulmonary involvement. Periodic home weights may aid in diuretic management, as dehydration is a common finding in affected patients.

Strategies utilized for COVID-19 management in the ACHD patient may include early detection and treatment of pneumonia, as well as use of bronchodilators for residual shortness of breath. The relationship between renin angiotensin system blockers and COVID-19, and the concern for increased risk of severe disease has been a subject of much debate, which has been clarified as non-significant by several large investigations [24,25]. On March 17, 2020, the ACC, American Heart Association and Heart Failure Society of America recommended the continuation of renin aldosterone angiotensin system antagonists for those who are currently prescribed these agents for indications which these agents are known to be beneficial, such as heart failure, hypertension, or ischemic heart disease [26].

INR may be impacted with any acute illness, especially for patients demonstrating reduced dietary intake or diarrhea, and therefore should be closely monitored for patients with COVID-19 who take VKAs (see considerations for transition to NOACs above). Of note, lopinavir-ritonavir, experimentally used in COVID-19 treatment, interacts with NOACs [27]. Thrombotic complications in patients with COVID-19 have emerged as an important issue during the pandemic [28]. Studies have found anticoagulation therapy in severe COVID-19 patients with markedly elevated D-dimer was associated with better outcomes [29,30]. Although patients with cyanosis and Fontan palliation have an inherent risk of thrombosis, a tailored approach with careful consideration of anticoagulation should be performed. Medication compliance for those already on anticoagulation should be reinforced.

5 Limitations

There is currently a gap in knowledge about the management and complications of the ACHD patient with COVID-19, and many COVID-19 publications are experiential. Further studies are underway in the ACHD field to address these gaps in literature.

6 Conclusion

This brief report depicts our local experience with ACHD patients during the COVID-19 global health crisis and how we adapted our program to accommodate their continuous cardiac needs while reducing their risk of exposure to COVID-19. As the rate of COVID-19 infections continues to fluctuate in various geographic locations in the coming months, ACHD programs must be prepared to adapt to offer continuous cardiac care largely at home and have strategies in place to manage patients with active COVID-19 infections. Larger investigations are currently underway to provide insight as to the effects of COVID-19 in this complex group of patients.

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References

1. Bansal, M. (2020). Cardiovascular disease and COVID-19. *Diabetes & Metabolic Syndrome*, 14(3), 247–250. DOI 10.1016/j.dsx.2020.03.013.
2. Tan, W., Aboulhosn, J. (2020). The cardiovascular burden of coronavirus disease 2019 (COVID-19) with a focus on congenital heart disease. *International Journal of Cardiology*, 309, 70–77. DOI 10.1016/j.ijcard.2020.03.063.
3. Stout, K. K., Daniels, C. J., Aboulhosn, J. A., Bozkurt, B., Broberg, C. S. et al. (2019). 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American college of cardiology/American heart association task force on clinical practice guidelines. *Journal of the American College of Cardiology*, 73(12), 1494–1563. DOI 10.1016/j.jacc.2018.08.1028.
4. Lin, M., Yuan, W., Huang, T., Zhang, H., Mai, J. et al. (2017). Clinical effectiveness of telemedicine for chronic heart failure: a systematic review and meta-analysis. *Journal of Investigative Medicine*, 65(5), 899–911. DOI 10.1136/jim-2016-000199.
5. Driggin, E., Madhavan, M. V., Chuich, T., Laracy, J., Biondi-Zoccai, G. et al. (2020). Cardiovascular considerations for patients, health care workers, and health systems during the coronavirus disease 2019 (COVID-19) pandemic. *Journal of American College of Cardiology*, 75(18), 2352–2371. DOI 10.1016/j.jacc.2020.03.031.
6. Chang, D., Saleh, M., Gabriels, J., Ismail, H., Goldner, B. et al. (2020). Inpatient use of ambulatory telemetry monitors for COVID-19 patients treated with hydroxychloroquine and/or azithromycin. *Journal of American College of Cardiology*, 75(23), 2992–2993. DOI 10.1016/j.jacc.2020.04.032.
7. FDA Website (2020). *Drug safety and availability*. <https://www.fda.gov/drugs/drug-safety-and-availability/fda-cautions-against-use-hydroxychloroquine-or-chloroquine-covid-19-outside-hospital-setting-or>
8. Chorin, E., Dai, M., Shulman, E., Wadhvani, L., Bar-Cohen, B. et al. (2020). The QT interval in patients with COVID-19 treated with hydroxychloroquine and azithromycin. *Nature Medicine*, 26, 808–809. DOI 10.1038/s41591-020-0888-2.
9. Rosenberg, E. S., Dufort, E. M., Udo, T., Wilberschied, L. A., Kumar, J. et al. (2020). Association of treatment with hydroxychloroquine or azithromycin with in-hospital mortality in patients with COVID-19 in New York State. *JAMA*, 323(24), 2493–2502. DOI 10.1001/jama.2020.8630.
10. Weinick, R. M., Burns, R. M., Mehrotra, A. (2010). Many emergency department visits could be managed at urgent care centers and retail clinics. *Health Affairs*, 29(9), 1610–1636. DOI 10.1377/hlthaff.2009.0748.
11. Hollander, J. E., Carr, B. G. (2020). Virtually perfect? Telemedicine for COVID-19. *New England Journal of Medicine*, 382(18), 1679–1681. DOI 10.1056/NEJMp2003539.

12. Harris, R. C., Fries, M. H., Boyle, A., Adeniji-Adele, H., Cherian, Z. et al. (2014). Multidisciplinary management of pregnancy in complex congenital heart disease: a model for coordination of care. *Congenital Heart Disease*, 9(6), E204–E211. DOI 10.1111/chd.12163.
13. Fernandes, S., Sanders, L. M. (2015). Patient-centered medical home for patients with complex congenital heart disease. *Current Opinion in Pediatrics*, 27(5), 581–586. DOI 10.1097/MOP.0000000000000258.
14. Barcellona, D., Fenu, L., Cornacchini, S., Marongiu, F. (2013). Telemedicine can improve the quality of oral anticoagulation using portable devices and self-testing at home. *Journal of Telemedicine and Telecare*, 19(6), 298–301. DOI 10.1177/1357633X13501764.
15. Holbrook, A., Schulman, S., Witt, D. M., Vandvik, O., Fish, J. et al. (2012). Evidence-based management of anticoagulant therapy. *Chest*, 141(2), e152S–e184S.
16. Yang, H., Veldtman, G. R., Bouma, B. J., Budts, W., Niwa, K. et al. (2019). Non-vitamin K antagonist oral anticoagulants in adults with a Fontan circulation are they safe. *Open Heart*, 6(1), 1–5. DOI 10.1136/openhrt-2018-000985.
17. Warnes, C. A. (2017). Adult congenital heart disease: the challenges of a lifetime. *European Heart Journal*, 38(26), 2041–2047. DOI 10.1093/eurheartj/ehw529.
18. Goyal, P., Choi, J. J., Pinheiro, L. C., Schenck, E. J., Chen, R. et al. (2020). Clinical characteristics of COVID-19 in New York City. *New England Journal of Medicine*, 382(24), 2372–2374. DOI 10.1056/NEJMc2010419.
19. Meng, X., Deng, Y., Dai, Z., Meng, Z. (2020). COVID-19 and anosmia: a review based on up-to-date knowledge. *American Journal of Otolaryngology*, 41(5), 102581. DOI 10.1016/j.amjoto.2020.102581.
20. Jackson, J. L., Leslie, C. E., Hondorp, S. N. (2018). Depressive and anxiety symptoms in adult congenital heart disease: prevalence, health impact and treatment. *Progress in Cardiovascular Disease*, 61(3–4), 294–299. DOI 10.1016/j.pcad.2018.07.015.
21. Duan, L., Zhu, G. (2020). Psychological interventions for people affected by the COVID-19 epicenter. *Lancet Psychiatry*, 7(4), 300–302. DOI 10.1016/S2215-0366(20)30073-0.
22. Zhou, X., Snoswell, C. L., Harding, L. E., Bambling, M., Edirippulige, S. et al. (2020). The role of telehealth in reducing the mental health burden from COVID-19. *Telemedicine Journal and E-Health: The Official Journal of the American Telemedicine Association*, 26(4), 377–379. DOI 10.1089/tmj.2020.0068.
23. Calcaterra, G., Crisafulli, A., Guccione, P., Di Salvo, G., Bassareo, P. P. (2019). Infective endocarditis triangle. Is it the time to revisit infective endocarditis susceptibility and indications for its antibiotic prophylaxis? *European Journal of Preventative Cardiology*, 26(16), 1771–1774. DOI 10.1177/2047487319856126.
24. Bavishi, C., Maddox, T. M., Messerli, F. H. (2020). Coronavirus disease 2019 (COVID-19) infection and renin angiotensin system blockers. *JAMA Cardiology*. DOI 10.1001/jamacardio.2020.1282.
25. Reynolds, H. R., Adhikari, S., Pulgarin, C., Troxel, A. B., Iturrate, E. et al. (2020). Renin-angiotensin-aldosterone system inhibitors and risk of COVID-19. *New England Journal of Medicine*, 382(25), 2441–2448. DOI 10.1056/NEJMoa2008975.
26. HFSA/ACC/AHA (2020). *HFSA/ACC/AHA statement addresses concerns re: using RAAS antagonists in COVID-19*. American College of Cardiology. <https://www.acc.org/latest-in-cardiology/articles/2020/03/17/08/59/hfsa-acc-aha-statement-addresses-concerns-re-using-raas-antagonists-in-covid-19>.
27. UpToDate (2020). *Lopinavir and ritonavir: drug information*. <https://www.uptodate.com/contents/lopinavir-and-ritonavir-drug-information>.
28. Giannis, D., Ziogas, I. A., Gianni, P. (2020). Coagulation disorders in coronavirus infected patients: COVID-19, SARS-CoV-1, MERS-CoV and lessons from the past. *Journal of Clinical Virology: The Official Publication of the Pan American Society for Clinical Virology*, 127, 1–4. DOI 10.1016/j.jcv.2020.104362.
29. Tang, N., Bai, H., Chen, X., Gong, J., Dengju, L. et al. (2020). Anticoagulant treatment is associated with decreased mortality in severe coronavirus disease 2019 patients with coagulopathy. *Journal of Thrombosis and Haemostasis*, 18(5), 1094–1099. DOI 10.1111/jth.14817.
30. Zhai, Z., Li, C., Chen, Y., Gerotziakas, G., Zhang, Z. et al. (2020). Prevention and treatment of venous thromboembolism associated with coronavirus disease 2019 infection: a consensus statement before guidelines. *Thrombosis Haemostasis*, 120(6), 937–948. DOI 10.1055/s-0040-1710019.