

CSANZ Position Statement on COVID-19 From the Paediatric and Congenital Council[☆]



Julian Ayer^{a,*}, Benjamin Anderson^b, Thomas L. Gentles^{c,**},
Rachael L. Cordina^d

^aThe Sydney Children's Hospital Network, Sydney, NSW, Australia

^bQueensland Paediatric Cardiac Service, Brisbane, Qld, Australia

^cStarship Children's Hospital, Auckland, New Zealand

^dRoyal Prince Alfred Hospital, Sydney, NSW, Australia

At the time of writing (25 May 2020), there have been nearly 4.4 million infections and 300,000 deaths worldwide related to COVID-19, an infection caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2). Australia (currently 6,900 infections and 98 deaths) and New Zealand (1,500 infections and 21 deaths) have thus far been less affected than other regions. Risk factors for more severe disease include older age and pre-existing cardiovascular disease. The purposes of this document from the Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand (CSANZ) are to: 1) To review the mechanisms for cardiac involvement in COVID-19, specifically as they may impact patients with childhood and adult congenital heart disease (CHD); 2) To review the impact of SARS-CoV-2 infection in the paediatric population; 3) To review available data on the risks related to COVID-19 for childhood heart disease and adult CHD; 4) To provide guidance for childhood heart disease and adult CHD units in our Australasian region to re-organise services during the pandemic, so as to protect a highly specialised workforce and yet continue to provide an essential service; and 5) To review risk reduction strategies for acquiring COVID-19 for patients with childhood heart disease or adult CHD. Eleven (11) recommendations relevant to the care of children with heart disease and adults with CHD to mitigate the impact of COVID-19 are highlighted through the document.

Keywords

COVID-19 • Childhood heart disease • Adult congenital heart disease • Pandemic response

Introduction

This Position Statement from the Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand (CSANZ) provides guidance for childhood heart disease and adult congenital heart disease (CHD) units in our region (Australasia) to understand the risks, mechanisms and impact of COVID-19 and to re-organise services during the current pandemic in order to protect a highly specialised

workforce and yet continue to provide an essential service. Eleven (11) recommendations relevant to the care of children with heart disease and adults with CHD to mitigate the impact of COVID-19 are highlighted through the document.

Mechanisms for Cardiovascular Involvement in COVID-19

COVID-19 predominantly affects the lungs causing an interstitial pneumonitis and acute respiratory distress

*Corresponding author at: The Sydney Children's Hospital Network, Sydney, NSW, Australia

**Corresponding author at: Starship Children's Hospital, Auckland, New Zealand, Email: TomG@adhb.govt.nz

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syndrome. However, multiple other organs, including the cardiovascular system, can become involved. Viral characteristics resulting in infection have been previously well reviewed [1]. Of particular note, severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) binds to the angiotensin converting enzyme 2 (ACE 2) receptor to enter host cells. The ACE 2 receptor is highly expressed in the lungs and cardiovascular tissue. Several factors related to infection have the potential of impacting the cardiovascular system including direct myocardial viral replication and damage, immune activation and cytokine storm, micro- and macrovascular dysfunction and thrombosis. A number of specific cardiovascular manifestations of severe COVID-19 infection warrant mention.

COVID-19 and myocardial injury and heart failure

Myocardial injury associated with COVID-19 has been suggested from a number of case reports and series [2]. Myocardial injury has been accompanied by elevation of serum troponin levels and left ventricular (LV) dysfunction. Not unexpectedly, evidence of myocardial injury appears to be more prevalent in fatal versus non-fatal COVID-19 infection. These manifestations may relate to direct viral-induced myocarditis and/or a broad systemic inflammatory response and supply/demand mismatch without acute atherothrombosis (type 2 myocardial infarction).

COVID-19 and arrhythmia

Early reports from China suggested an association of severe COVID-19 infection with arrhythmia [3,4]. Factors that may promote arrhythmia with severe COVID-19 infection include metabolic derangement, myocarditis/myocardial inflammation and sympathetic activation.

COVID-19 and coagulation abnormalities

Coagulation abnormalities including disseminated intravascular coagulation (DIC), systemic and arterial thrombosis and pulmonary thromboembolism have been reported in severe COVID-19 infection [5–7].

The impacts of these effects of COVID-19 infection on patients with childhood or adult congenital heart disease, particularly those more at risk of ventricular dysfunction, arrhythmia and thrombosis, are as yet unknown.

Impact of SARS-CoV-2 Infection in Paediatric Populations

SARS-CoV-2 has been reported to infect children, although current literature suggests both that paediatric patients make up only a small percentage of the total number of documented cases and that the disease process tends to be less severe than reported in the adult population [8].

Demographics

Data from countries heavily impacted by the pandemic have consistently reported low percentages of children in the total numbers of confirmed cases. These range from

1% in China, Italy and Iran, up to 5% in the United States, with 2.4% reported by the World Health Organization (WHO)-China Joint Mission on Coronavirus. The median age of reported paediatric cases is around 7 years, with no significant difference in sex distribution (approximately 60% in boys). Rates of paediatric infection in Australia are currently very low (at time of writing, about 1% of cases in Queensland, 3% in New Zealand, and 4% in New South Wales, for example).

Severity of illness

The severity of the illness appears to be milder in the paediatric population than reported in adults [8–10]. Of those children infected, the illness was asymptomatic, mild or moderate in over 90% of cases. Mild symptoms (of upper respiratory tract infection [URTI]) were the most commonly reported (around 50%), with moderate symptoms (fever, cough, consistent with pneumonia) reported in 40%. Severe symptoms, manifest with hypoxia (saturation <92%) were present in around 2.5% of cases and critical symptoms with respiratory failure and end organ damage or intensive care unit (ICU) admission in 0.4% to 1.7% of cases. Severe illness has been reported to be more likely in the younger age groups (<1 yr=10%; 1–5 yr=7%; 6–15 yr=4%; >16 yr=3%). Fever was present at some time during the illness in 42% but was often less than 38° (in around 60% of cases). Asymptomatic infection has been reported in varying numbers of paediatric patients, ranging from 4–16% of confirmed positive cases.

Mode of infection

Of those children exposed to an index case with known or suspected SARS-CoV-2 infection, 12% subsequently tested positive. Ninety per cent (90%) of these cases were identified through contact tracing of a symptomatic adult as part of a family cluster. Small case series suggest the main source of infection in children is transmission from an adult family member, based on the infected children having developed symptoms after exposure to the index adult case [11].

Paediatric inflammatory multi-system syndrome temporally associated with SARS-CoV-2 (PIMS-TS)

Recently, a small number of cases of a multi-system inflammatory syndrome associated with paediatric COVID-19 infection have been described [12]. This syndrome, marked by fever, inflammation and poor function in one or more organ, has features overlapping with Kawasaki disease (KD), toxic shock and macrophage activation syndromes. Transient ventricular dysfunction requiring intensive care admission, inotropic and/or extracorporeal membrane oxygenation (ECMO) support but with rapid recovery has recently been reported as part of PIMS-TS [13]. Children with these features warrant close review of clinical and inflammatory markers, echocardiography and standard supportive management. Intravenous immunoglobulin (IVIg) and aspirin could be considered if fulfilling criteria for KD.

Recommendation 1: Clinicians suspecting a case of paediatric multi-system inflammatory syndrome associated with COVID-19 should consult promptly with a paediatric infectious disease, rheumatology, cardiology and/or critical care physician.

Recommendation 2: A system of regional surveillance of COVID-19 associated paediatric multi-system inflammatory syndrome should be developed.

Cardiovascular Risk Factors for Severe COVID-19 Infection

At the time of writing, there is no evidence to support a harmful effect of ACE inhibition in COVID-19 infection [14,15], and a recent brief report from the UK Clinical Practice Research Datalink suggests that there may be a dose-related protective effect [16]. Given the potential benefit of these medications they should be continued.

Recommendation 3: ACE inhibitors in children with heart disease and adults with chronic heart disease (CHD) should be continued during the COVID-19 pandemic.

Very little data exists on the specific risks for severe COVID-19 infection in childhood heart disease and adults with CHD. In a description of 485 adult CHD cases with clinical COVID-19 infections from Italy, 70% were asymptomatic, 20% had specific symptoms and 10% non-specific symptoms. No patients had severe disease and only two were hospitalised (The Sommerville Foundation Live Webinar; <https://www.livemediacom/achdcovid19>). Risk factors for severe COVID-19 appeared to relate less to the underlying cardiac condition and more to other comorbidities.

Nonetheless, using the precautionary principle, the following groups of patients could represent those at higher risk for severe COVID-19 infection: Fontan circulation or single ventricle palliation, chronic cyanosis, significant ventricular dysfunction, pulmonary hypertension, those with impaired immunity (22q11 deletion, asplenia) or other comorbidities (renal failure, lung disease). These “higher risk” groups are similar to those identified in a recent British Congenital Cardiac Association newsletter (https://www.bcca-uk.org/pages/news_box.asp?NewsID=19495710).

Maintaining Access to Health Care

During the COVID-19 pandemic, it is important that patients with childhood heart disease and adult congenital heart disease maintain their access to health care.

Access to medical assessment and treatment for non-COVID conditions has been reduced during the pandemic. This is the case in all areas of medical care from primary health to the specialist hospital.

Notwithstanding that some patients and families are reluctant to visit medical facilities for fear of infection, the major contributor is a reduction in service provision. This has occurred so that resources can be diverted to the treatment of COVID-positive patients, and to reduce the risk of COVID-19 spread within the hospital environment through social distancing and other protective strategies. In addition, the usual level of community supervision may be impacted either because there is a reluctance of health care workers to visit homes, or because local community health services have curtailed this kind of service. This has the potential to adversely impact not only the health and wellbeing of fragile patients but also the delivery of secondary rheumatic fever prevention and other community-based interventions. Treatment may also be delayed because of minor respiratory symptoms which are especially common in children.

Recommendation 4: Paediatric Cardiology and Adult Congenital Cardiology Services should aim to ensure that adequate services are provided to facilitate telehealth consultations to minimise patient exposure related to travel, clinic and hospital visits when clinically appropriate. This type of consultation is especially relevant to those living remotely.

Recommendation 5: Those waiting and/or deferred for investigation and treatment will benefit from frequent review of acuity. While this can be aided by remote telemedicine assessment, it should be recognised that this is a risk mitigation, rather than risk elimination, strategy.

Recommendation 6: Clinicians and health services need to ensure, when possible, that patients who have chronic heart disease (CHD) or childhood heart disease are not deprived of acceptable levels of care due to diversion of resources to COVID pathways. This is especially relevant for patients who are socially disadvantaged and/or live remotely where assessment may be beyond the expertise of local medical teams. Resources need to be ring-fenced so that time-critical investigation and treatment can proceed.

Recommendation 7: Specifically, for children and adults with a history of acute rheumatic fever and/or rheumatic heart disease, continued prophylaxis with 3–4 weekly benzyl penicillin injections remains critically important.

Paediatric cardiac units and adult CHD services in the region are dependent on small numbers of key personnel. In some instances—particularly cardiac surgery—absence of these personnel because of exposure to, or illness from,

COVID-19 will have a profound operational impact. This is also the case for those staff with a less unique skill mix if they are absent in large numbers.

Recommendation 8: Patients and families should be screened prior to hospital admission or clinic visits to ensure that they do not have symptoms attributable to COVID-19 infection, have not had contact with someone who is infected, and are aware that if they develop these symptoms prior to the visit they should not attend. In accordance with local health department recommendations, consideration should be given to COVID testing of all admissions and clinic visits, if community infection rates of COVID-19 are high.

Recommendation 9: Personal protection equipment (PPE) should be available and utilised as per local hospital protocols.

Recommendation 10: Appropriately timed influenza vaccine is especially important during the COVID-19 pandemic as co-infection may increase the risk for serious respiratory complications. Patients and family members over the age of 6 months of age should be encouraged to be vaccinated unless they have a contraindication.

Recommendation 11: Provision of up-to-date information to families and patients is important. This should be readily available electronically, updated regularly, and relevant to the local environment.

Reducing the Risk of Acquiring COVID-19

Although data are lacking, it is likely that older patients with complex CHD are at increased risk for COVID-19 related complications. As outlined above, if community COVID-19 rates are high enough to pose a significant risk, visits to clinics and hospitals for investigations, assessments and procedures should be delayed if deemed non-urgent. Consultations conducted via videoconferencing or telephone may suffice for stable patients.

It is important that the risks for delaying investigation and treatment do not outweigh the potential risk for COVID-19 complications and patients should be encouraged to access appropriate care when needed. Parents, patients, and families are likely to feel especially vulnerable during the pandemic and may need additional reassurance by clinical care teams.

Patients and family should be counselled about general measures such as hand hygiene, respiratory hygiene, and social distancing, according to current regional

recommendations that may vary according to rates of community transmission. Low risk patients and their families should be encouraged to follow local guidelines to reduce the risk for infection including recommendations for work and school attendance. The decision to work or study from home for higher risk patients and family members may be more challenging and depends upon family resources, logistics, current community transmission rates and individual risk factors. Data regarding the efficacy of facial mask use in public are controversial. Physicians may consider recommending face covers in especially high-risk patients and situations.

Conclusion

The COVID-19 pandemic has been and will continue to be a great challenge to health care systems across the globe. To date, little information is available to indicate that those with childhood heart disease and adult CHD are at a higher risk of severe COVID-19 infection. However, until further data is available, those patients with more severe forms of heart disease should pay particular attention to measures to avoid COVID-19, such as social distancing and hand hygiene. The small but highly specialised teams comprising the childhood and adult CHD workforce will require reorganisation to ensure continued high-level service provision in the face of likely health worker infection. These services need to ensure that systems are in place to facilitate the continued timely provision of cardiac care to those patients who need it during the COVID-19 pandemic.

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