

## Management Congenital Heart Disease Surgery during COVID-19 : A Review Article

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### Abstract

Congenital heart disease is a form of heart abnormality that has been acquired since the newborn. The clinical course of this disorder varies from mild to severe. In mild forms, there are often no symptoms, and no abnormalities are found on clinical examination. Whereas in severe CHD, symptoms have been visible since birth and require immediate action. Generally, the management of congenital heart disease includes non-surgical management and surgical management. Non-surgical management includes medical management and interventional cardiology. Medical management is generally secondary as a result of complications from heart disease itself or due to other accompanying disorders. In this case, the goal of medical therapy is to relieve symptoms and signs in addition to preparing for surgery. The duration and method of administration of drugs depend on the type of disease at hand.

The severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), which led to the coronavirus disease 2019 (COVID-19) pandemic, was initially reported in Wuhan, China in December, 2019. The rapid rise in the number of cases worldwide led to hospitals struggling to cope with the sudden influx of patients. This has had a ripple effect on other parts of health care as manpower and supplies needed to be reallocated. Within cardiology, this has led to outpatient appointments and elective surgeries being reduced and/or postponed. COVID-19 appears to have a complicated relationship with cardiovascular system, as studies have suggested cardiovascular diseases increase disease severity and mortality rates in those who are infected. However, the virus has also been shown to cause cardiovascular complications such as acute myocardial injury, heart failure, and arrhythmia.

Coronavirus may also cause myocardial injury via the cytokine storm that occurs in response to a possible large immune response during the infection. Cardiac involvement such as right ventricular failure and congestion can either be a result of respiratory distress or direct cardiac injury caused by the virus, as suggested by the raised cardiac troponin I in critical patients compared to non-critical patients.

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### Introduction

Congenital heart disease is a form of heart abnormality that has been acquired since the newborn. The clinical course of this disorder

varies from mild to severe. In mild forms, there are often no symptoms, and no abnormalities are found on clinical examination. Whereas in severe CHD, symptoms have been visible since birth and require immediate action. With the development of technology, especially echocardiography, many heart abnormalities that previously could not be detected by physical examination and usual support, ECG, radiology using this tool can be detected easily<sup>1</sup>.

In most cases, the cause of CHD is unknown. Various types of drugs, maternal

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diseases, exposure to X-rays, are thought to be exogenous causes of congenital heart disease. Rubella disease suffered by the mother in early pregnancy can cause CHD in the baby. In addition to exogenous factors, there are also endogenous factors associated with the incidence of CHD. Various types of genetic diseases and certain syndromes are closely related to the incidence of CHD such as Down syndrome, Turner syndrome, and others<sup>1,2</sup>.

Congenital heart disease (CHD) is the most common and global inborn defect, affects approximately 1% of live births globally<sup>3</sup>. CHD is a term for a range of birth defects that have varying levels of severity depending on the type. Different types of CHDs include coarctation of the aorta (CoA), atrial septal defects (ASDs), and ventricular septal defects (VSDs) (NHS). Studies have suggested that patients with CHD may be at an increased risk of complications, and thus poorer outcomes, if they acquire COVID-19. Although some data have shown no such correlation<sup>3</sup>.

Generally, the management of congenital heart disease includes non-surgical management and surgical management. Non-surgical management includes medical management and interventional cardiology. Medical management is generally secondary as a result of complications from heart disease itself or due to other accompanying disorders. In this case, the goal of medical therapy is to relieve symptoms and signs in addition to preparing for surgery. The duration and method of administration of drugs depend on the type of disease at hand<sup>1,3</sup>.

## Discussion

The severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), which led to the coronavirus disease 2019 (COVID-19) pandemic, was initially reported in Wuhan, China in December, 2019. The rapid rise in the number of cases worldwide led to hospitals struggling to cope with the sudden influx of patients. This has had a ripple effect on other parts of health care as manpower and supplies needed to be reallocated. Within cardiology, this has led to outpatient appointments and elective surgeries being reduced and/or postponed. COVID-19 appears to have a complicated relationship with cardiovascular system, as studies have suggested cardiovascular diseases increase

disease severity and mortality rates in those who are infected. However, the virus has also been shown to cause cardiovascular complications such as acute myocardial injury, heart failure, and arrhythmia<sup>4</sup>.

CHD require continuing care, particularly amongst newborns and infants who often require surgery during a narrow window of time to avoid death and provide for optimal outcomes. Crisis management strategies for congenital heart disease have recently been published (Stephens, et al). The strategies deal with many aspects such as social exposure and distancing, triage, timing and role of the surgeon. However, social distancing can be particularly challenging for our patients, given their age and family structure. In addition, asymptomatic children can be carriers and especially for younger it is difficult to keep masks and maintain social distancing. For this reason testing should be performed on every patient, even asymptomatic patients, as well as their parents. In this period, cooperation between surgeons and cardiologists is even more important to evaluate hospital resource utilization, clinical status of the patient and risk of delaying surgery, comorbidities and complexity of the procedure and risk of exposure for the patient, family and healthcare staff<sup>5</sup>.

In addition, Levy and colleagues try in a short communication to answer the most frequent questions for pediatric patients with CHD (Levy, et al). However, there are few established concepts: (1) the best test for SARS-CoV2 in the peri-operative setting is a PCR of respiratory secretions. Serology serum testing for antibodies (IgG) will demonstrate prior exposure (or maternal status for neonates) rather than active illness, so is less useful in a peri-operative setting; (2) CT scans should not be used to screen for or diagnose pediatric COVID-19. CT scans should be reserved for other clinical indications based on symptoms; (3) All patients entering the hospital or clinic should be screened for symptoms suggesting COVID-19 and for test screening performed routinely; (4) for infants born to COVID positive mothers should be reasonable to separate him from the mother if will need cardiac surgery to try avoid post-natal infection. In fact, there is minimal evidence of placental vertical transmission. It may also be reasonable to do serial testing on the infant, but there is no consensus on the correct timing (Schwartz, et al); (5) surgery should be

scheduled with advice from a multidisciplinary team of experts including cardiac medical, cardiac surgical, and infectious diseases as indicated. If prudent, surgery should be delayed until the patient's symptoms have improved and/or testing has been repeated (often after 14 days) and is negative<sup>6</sup>.

The use of lung ultrasound in the pediatric population could be very useful for screening and management, as already we have demonstrated in the management of children with CHD (Cantinoti et al; cantinoti, et al). The adoption of lung ultrasound to monitor lung disease during the COVID-19 outbreak may reduce the number of chest X-ray, limiting dangerous radiation cumulative exposure (cantinoti, et al). Lung ultrasound is an easy, cheap and fast technique that can be repeated at patient bed. While in adult setting (especially in overweight patients) image quality of lung ultrasound may be suboptimal, in the small thorax of children image quality is usually extremely high. Furthermore in the small thorax of the children, lung examination is faster and a complete and accurate examination of all lung sectors may be achieved in a few minutes<sup>7</sup>.

Even though COVID-19 infection in childhood is less common and with milder symptoms than when occurring in adult patients, it is not without the risk of cardiac involvement, especially in the patients with a background of congenital heart disease. In newborns and children, previous cardiac surgery is related with the risk of a more severe form of the disease, being admitted to intensive care unit, and needing intubation as well as mechanical ventilation<sup>8</sup>.

One study reported two patients with unspecified repaired CHD, aged 8 months and 1 year, respectively. These two cases showed the most severe disease progression, as they developed impaired renal function, with high levels of lactate dehydrogenase and impaired coagulative function. One of the reported patients also had a suspected hereditary metabolic disease and malnutrition. They both developed ARDS and were admitted to ICU, requiring mechanical ventilation<sup>9</sup>.

In regard to ACHD, Sabatino et al conducted a nationwide survey in Italy that included patients with CHD who either were suspected of having COVID-19 or confirmed. Interestingly, all patients with poor outcomes

were confirmed COVID-19 positive, as opposed to the mild disease progression that was experienced by those only suspected of being COVID-19 positive. They suffered from a range of different CHDs including TGA and VSDs. One of them required continuous positive airway pressure (CPAP) therapy, whilst another was put on extracorporeal membrane oxygenation (ECMO) therapy and ended up in ICU. The majority of these patients experienced complications including heart failure (55%), stroke, and arrhythmias. Arrhythmias are frequently seen in CHD-COVID-19-positive patients with palpitations accounting for 22% of the presenting complaints and this often requires intensive care treatment<sup>10</sup>.

A study outlined the categories of children with CHD that would be at increased risk of severe illness if they acquired a COVID-19 infection. These included patients with a large VSD, "single heart physiology" such as hypoplastic left heart syndrome and tricuspid atresia, and patients who had an upcoming surgical procedure or had already undergone one, including the Fontan procedure<sup>11,12</sup>.

Whilst the aetiology of cardiovascular complications due to SARS-Cov-2 infection is not clear, it has been proposed that ACE2 receptors could be used as a mode of entry into myocytes and cause myocardial injury through various mechanisms. 2019-nCoV gains access into the cells by binding to the same receptor, angiotensin-converting enzyme II (ACE2), as SARS-CoV<sup>13,14</sup>.

SARS-CoV-2 downregulates the expression of ACE2 once inside the cells in which ACE2 is responsible for converting angiotensin II to angiotensin 1–7. Angiotensin II has a pro-inflammatory and pro-fibrotic role as well as being a vasoconstrictor, whereas angiotensin 1–7 play an important anti-inflammatory and antioxidant role as well as causing mild vasodilation, protecting the heart and lungs from injury. Angiotensin II levels are found to be notably increased in the plasma of those infected with SARS-CoV-2 whilst high ACE2 levels were seen in those less affected by the virus and these levels reportedly decrease with age. Decreasing levels of ACE2 may explain why the elderly are at a higher risk of severe illness from COVID-19 compared to the paediatric population<sup>15,16</sup>.

High ACE2 levels seen in the younger

population can be protective as it leads to increased conversion of angiotensin II to angiotensin 1–7 to counteract any vasoconstriction, downregulate cytokines, leukocytes, and fibrosis. On the other hand, in those with low levels of ACE2, angiotensin II accumulates and can lead to pro-inflammatory effects resulting in severe cases. Another factor that could contribute to the reduced cases and a better prognosis in the paediatric population may be the absence of co-morbidities and fewer risk factors such as smoking and obesity<sup>7,17,18</sup>.

### Management of CHD disease

Midline sternotomy incision, deepened layer by layer, bleeding is stopped, the sternum is opened with a sternal saw, the bleeding is stopped (if necessary with bone wax), attach a sternum retractor. The thymus is removed or removed, the pericardium is opened and fixed/stitched to the wound margin with silk 3.0 in several places. Identification of the Inominata or PLSVC . vein. Identification of PDA. Preparation of cannulation, heparin is given 3 mg/kgBW bolus IV, Sutures cannulate the aorta, SVC, and IVC with 4 non-absorbable braided 3.0/4.0 sutures or 4 monofilament 5.0 threads. Check ACT and AGD. If ACT > 200, the aorta is cannulated, SVC and IVC are connected to the CPB machine. Cannula fixed with 2.0 silk thread 1 piece. The CPB engine is run when ACT400, the temperature is lowered gradually to 32-34 degrees Celsius<sup>6,19,20</sup>.

SVC and IVC were totalo-occlusion using cotton tape. If PDA is found, PDA is ligated with 1 non-absorbable braided 2.0. Placed ulcardioplegia or IV Catheterno.14/16 on the aortic root and connected to the line cardioplegia of perfusionis. The aortic cross clamp is placed on the ascending aorta before the aortic cannula, cold cardioplegic fluid is infused into the aortic root. The RA is opened parallel to the atrioventricular sulcus. The edges are fixed with silk 3.0.(4.0). Identify ASD, pulmonary veins, SVC and IVC estuaries, coronary sinus (unroofed or not), mitral valve (make sure there is no cortriatiatum) and tricuspid valve. If there is a PAPVD, re-routing ASDClosure At the ASDP Primum, carefully evaluate the mitral valve if present cleft and regurgitation performed MV repair with sutures on the cleft with polypropylene 5.0/6.0 interrupted 4-5 pieces<sup>6,21</sup>.

ASD Closure with or without patch pericardium with using 5.0 or 4.0 . polypropylene.

Evaluation of residual ASD, if any, closed with thread sutures polypropylene 5.0 or 4.0. Body temperature is increased gradually. Deairing of the left heart, rapid lung development, position Trendelenburg patient. Klemscrossaorta is released, the heart beats spontaneously. PAline is entered directly to PA in ASD casewith severe PH (MPAP > 2/3 systemic). RA is closed (MPA in cases of ASD with PHsevere)with 5.0 or 6.0 polypropylene yarn while deairing right heart. Total occlusion SVC and IVC removed<sup>5,22</sup>.

### Conclusion

Coronavirus may also cause myocardial injury via the cytokine storm that occurs in response to a possible large immune response during the infection. Cardiac involvement such as right ventricular failure and congestion can either be a result of respiratory distress or direct cardiac injury caused by the virus, as suggested by the raised cardiac troponin I in critical patients compared to non-critical patients.

### Declaration of Interest

The authors report no conflict of interest.

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