COVID-19 and genetic heart disease

A summary of the Cardiac Society of Australia and New Zealand guidelines

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Genetic heart diseases are relatively rare in the population; however, they have very serious, potentially life-threatening consequences. Data to inform best practice for patients with genetic heart disease in light of the current severe acute respiratory syndrome coronavirus pandemic are lacking. Identifying and regularly reviewing areas of consensus among leading specialists is essential for protecting patients who are at increased risk of morbidity and mortality during the pandemic.

Sudden cardiac death (SCD) is defined as a death, presumed to be cardiac in origin, in an apparently healthy subject occurring within an hour of the onset of symptoms or unwitnessed deaths where the decedent was well 12 to 24 hours before death.[1] SCD has a prevalence of 1.3 cases per 100,000 persons in adults under the age of 35 years in Australia and New Zealand.[2] A significant proportion of SCD in the young may be precipitated by underlying genetic heart diseases, encompassing both inherited cardiomyopathies and channelopathies in genes responsible for sodium-, potassium- and calcium- channel function.[2]

The severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) is responsible for the coronavirus disease 2019 (COVID-19) pandemic.[3] It is well documented that pre-existing cardiovascular disease increases morbidity and mortality among patients with COVID-19.[3] Accordingly, the Cardiac Society of Australia and New Zealand (CSANZ) released a set of recommendations pertaining to COVID-19 in patients with genetic heart disease, which are summarised below.[4]

Familial cardiomyopathies

- People who are gene carriers for a familial cardiomyopathy variant but who do not have clinical expression of the disease are possibly at risk and should be vigilant with preventative measures (e.g. social distancing, hand washing).[4]
- The CSANZ guidelines recommend that patients with severe left or right ventricular dysfunction or with symptomatic left or right ventricular failure are at highest risk, and therefore should take substantive efforts to minimise risk of exposure

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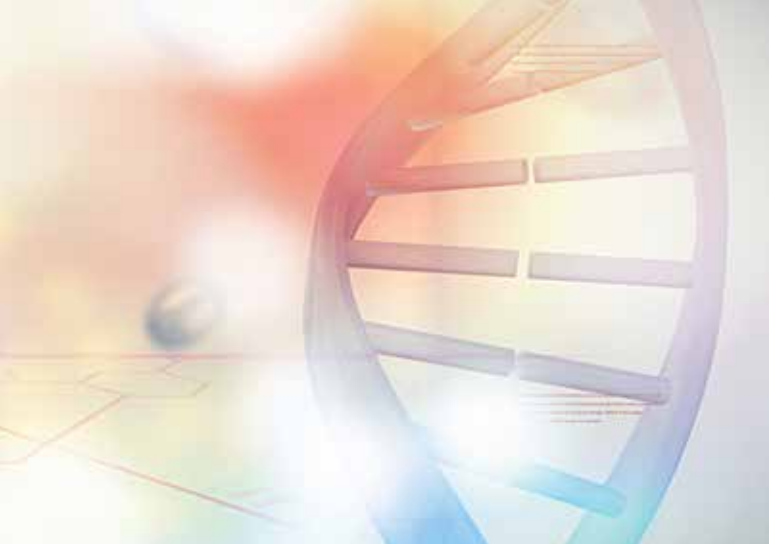

to COVID-19 (e.g. self-isolation, social distancing and hand washing).[4]
- Patients should continue their regular heart failure therapies where possible.[4]

Brugada syndrome (BrS)

- BrS is an inherited arrhythmia syndrome with a prevalence of 1:2000, most commonly due to genetic mutations in the sodium channel.[5]
- Patients with BrS are at risk of ventricular arrhythmias if they develop fever (over 38°C).
- The CSANZ guidelines recommend that patients with fever of more than 38°C should be treated aggressively with paracetamol and seek medical attention.[4]
- Patients with BrS who have a fever that is unresponsive to antipyretics should seek urgent specialist cardiology advice as they may require more intensive monitoring.[4] This is especially important for children with BrS.

Long QT syndrome (LQTS)

- LQTS is an inherited arrhythmia syndrome characterised by the presence of a prolonged QT interval (corrected for heart rate [QTc]) in the absence of secondary causes for a prolonged QTc interval such as drugs or electrolyte disturbances.[5]
- No current data suggest patients with LQTS are at increased risk from COVID-19 infection.
- The CSANZ guidelines recommend all beta-blockers are continued in patients with LQTS during their illness.[4]
- CSANZ guidelines also recommend exercising great caution when using pharmacological therapies for COVID-19 such as hydroxychloroquine, azithromycin and ritonavir, as these are known to prolong the QT interval even in patients without congenital LQTS.[4]
- Both the CSANZ and Heart Rhythm Society (HRS) guidelines support regular QT monitoring for all patients with COVID-19 in whom hydroxychloroquine combined antiviral drugs are started.[4,6]
- In patients with LQTS with significant gastrointestinal complications of COVID-19 (e.g. diarrhoea), CSANZ and HRS guidelines recommend close electrolyte monitoring (particularly potassium and magnesium) and prompt electrolyte replacement as required.[4,6]
- If the QTc interval is consistently greater than 500 ms, CSANZ and HRS guidelines recommend consultation with a cardiogenetics expert or electrophysiologist for guidance on further management to minimise risk of torsades de pointes.[4,6]

Catecholaminergic polymorphic ventricular tachycardia (CPVT)

- CPVT is a rare, inherited arrhythmia syndrome characterised by adrenergically stimulated ventricular tachycardia, typically through physical exertion, in individuals with a structurally normal heart.[5]
- There are no data to suggest patients with CPVT are at increased risk from COVID-19 infection.
- CSANZ guidelines recommend patients with CPVT continue their regular cardiac medications.[4]

Hospital admission

It should be noted that if a patient with a known genetic heart disease is admitted to hospital or intensive care or is started on novel therapies for COVID-19, a specialist cardiologist or one with suitable sub-specialty expertise should be consulted during this time.

Conclusion

Patients with a genetic heart disease who develop COVID-19 should be managed in consultation with cardiologists with subspecialised expertise to minimise their chance of life-threatening arrhythmic complications. CT

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