



CONSENSUS STATEMENT FOR PATIENTS WITH GENETIC HEART DISEASE AND COVID-19

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We must acknowledge that this is a newly discovered virus, and genetic heart diseases are relatively rare in the population. Therefore, there are few scientific data to inform best practice on this subject thus far. However, we have identified areas of consensus among specialists from the CSANZ cardiovascular genetics council with regards to managing patients with genetic heart disease in this current COVID-19 pandemic.

GENERAL RECOMMENDATIONS

- Patients with pre-existing structural heart disease are at increased risk of complications from COVID-19 and therefore we recommend all patients follow government recommendations to help minimise exposure and prevent infection (e.g. social distancing, hand washing)
- Patients should continue all of their cardiac medications including beta-blockers, calcium-channel blockers, ACE-Inhibitors (ACE-I), Angiotensin II Receptor Blockers (ARBs), anticoagulation etc. There have been some reports with conflicting advice with regards to ACE-I and ARBs. The concerns are theoretical regarding COVID-19 accessing cells via the ACE-2 receptors potentially leading to worse outcomes, but currently there is no strong evidence to substantiate this. Of greater concern is haemodynamic instability, and the risk of ventricular dysfunction due to sudden transition from one medication to another at a time of potential myocardial stress. A number of international cardiac societies have recommended continuation of a patient's current medications at present^{1,2}. We also support continuation of current medications, especially in the setting of impaired ventricular function and cardiomyopathy.
- We recommend all our patients have their annual influenza vaccination as soon as possible to reduce the chance of having seasonal influenza and COVID-19 infection at the same time.
- We recommend that if a patient with a known genetic heart disease is admitted to ICU, or is to be commenced on novel or atypical therapies (such as hydroxychloroquine), a specialist cardiologist, ideally their regular cardiologist or one with suitable sub-speciality expertise, should be consulted.

SPECIFIC RECOMMENDATIONS:

1) Cardiomyopathies

- People who are gene carriers for a familial cardiomyopathy variant, but without clinical expression of the disease are possibly at risk and should be vigilant with preventative measures.
- A patient's level of risk may be dependent on the degree of clinical expression. Those with severe ventricular dysfunction (of either the left or right ventricle) or with symptomatic left or right ventricular failure, should be considered at highest risk, and therefore take great efforts to minimise risk of exposure to COVID-19 (e.g. self-isolation).
- The management of patients who are unwell with COVID-19 and have an underlying cardiomyopathy should include specialist cardiology advice.

2) Brugada Syndrome

- Patients with Brugada syndrome are at risk of arrhythmia with high fever. Therefore, we recommend patients treat fever (>38C) aggressively with paracetamol and seek medical attention.
- BrS patients with fever that is unresponsive to paracetamol should seek urgent specialist cardiology advice as they may require more intensive monitoring.
- Children with BrS
 - As above, fever should be managed aggressively and medical attention sought in children with a type 1 Brugada ECG, or children of a parent with *SCN5A*-mediated Brugada syndrome unless they are known to be gene negative.
 - Preliminary evidence suggests that children of a parent affected with Brugada syndrome, in whom *SCN5A* pathogenic variants have been excluded, and where the child has a normal baseline ECG, are at much lower risk from fever than previously feared. Many of these non-*SCN5A* families have been advised to bring their children to hospital with a fever to see if the Brugada ECG signature manifests. In such children, during this pandemic, to avoid exposure to COVID-19 and reduce stress on hospitals, fever should be managed at home as usual and children who are well should not be brought into to the Hospital/ Emergency Department just for an ECG.

3) Long QT syndrome

- There are no data to suggest patients with LQTS are at increased risk from COVID-19 infection.

- We recommend all betablockers are continued in patients with LQTS during their illness.
- Some experimental therapies for COVID-19 (such as hydroxychloroquine, azithromycin and ritonavir) are known to prolong the QT interval, and may cause acquired LQTS even in patients without congenital LQTS; therefore, we advise great caution in use of these medications.³ We recommend that patients and their treating physicians consult www.crediblemeds.org prior to commencing any new medications. We support the proposed flow chart by Wu et al, *Heart Rhythm* 2020.⁴ Key points as below:
 - QT monitoring for all patients commenced on hydroxychloroquine or when combining anti-viral drugs for COVID-19
 - Avoid the use of more than one medication which prolongs the QT interval as far as is possible
 - If the QTc is consistently >500ms we recommend consultation with a cardiogenetics expert or electrophysiologist for guidance on further management to minimise risk of Torsade de Pointes.
- In patients with LQTS with significant gastrointestinal complications of COVID-19 (e.g. diarrhoea), we recommend their electrolytes (especially potassium) are checked and replaced as needed. We recommend a serum potassium at the higher end of the normal range (i.e. above 4 mmol/L)

4) **Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)**

- There are no data to suggest patients with CPVT are at increased risk from COVID-19 infection.
- We recommend CPVT patients continue their regular cardiac medications.
- We recommend avoidance of epinephrine in the setting of a VT/VF arrest where possible. We recognise this is likely to be the only situation where epinephrine is contraindicated in the setting of a cardiac arrest.

OTHER RECOMMENDATIONS FOR GENETIC HEART DISEASE PATIENTS AND THEIR GPs

- There is some suggestion NSAIDS may worsen the respiratory course of COVID-19 and we recommend paracetamol is used preferentially to manage fever.
- We understand many of our patients and their families are feeling worried or anxious about their risk of poor outcomes given their underlying heart disease. We recommend individuals seek additional support and information on coping in a crisis, and seek professional assistance as required (e.g. www.beyondblue.org.au,: www.psychology.org.au).

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