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# Rheumatic heart disease and COVID-19

## Critical considerations for patients, providers, and health systems dealing with rheumatic heart disease during and post-pandemic

Rheumatic heart disease (RHD) affects >39 million persons around the world, with the highest prevalence in low-resource populations with constrained health systems.<sup>1</sup> It is a sequel of rheumatic fever which occurs after a single or multiple episodes of sore throat due to group A beta-haemolytic streptococci. Secondary prophylaxis, consisting of intramuscular penicillin injections every 28 days, is the cornerstone of therapy for patients with RHD, protecting them from further streptococcal sore throat infections, recurrent rheumatic fever, and RHD progression. The COVID-19 pandemic presents a significant challenge for these patients, who are likely to encounter interruptions in secondary prophylaxis and access to care, with potentially dire consequences. Here, we address the direct and indirect risks that the COVID-19 pandemic poses to those living with RHD, in particular those living in low-resource environments, and provide expert guidance to support patients, providers, and policy makers during this global crisis.

### The direct risk of COVID-19 infection to people living with RHD

The World Heart Federation lists RHD as a risk factor for severe COVID disease 'unique to low-income countries'.<sup>2</sup> Although the magnitude of the direct risk is not known, this risk is likely to be higher in those living with more severe forms of valvular heart disease and highest among patients with pulmonary hypertension and those with heart failure, requiring medication to improve cardiac function.<sup>3</sup> Individual risk of death may be further augmented by resource constraints in low- and middle-income countries (LMICs) where most patients with RHD live, and where there is the least capacity to respond.

Patients living with RHD should, to the best of their ability, follow the basic infection control recommendations provided by the World Health Organization (WHO) as well as their local and national governments. As RHD is dynamic, we recommend that when possible, patients living with RHD should keep scheduled medical appointments to monitor for progression and cardiovascular complications, and should be given 'essential' status to travel to these appointments under local regulations. We recommend that patients continue to take all prescribed cardiac medications, including those classified as angiotensin-converting enzyme inhibitors (ACEIs) and angiotensin II receptor blockers (ARBs), due to a lack of data supporting COVID-specific risk of these therapies. In addition, patients with prosthetic heart valves on warfarin and requiring regular international normalized ratio (INR) checks will find this time particularly challenging to access regular review, where clinics and even hospitals are curtailing normal services.

When considering direct risk, it is also of critical importance to remember that only a small fraction of those living with RHD in LMICs have been diagnosed. In the context of the COVID-19 pandemic, this represents a group of potentially vulnerable persons who will not be

aware of their increased risk. Countries with endemic RHD who are facing the COVID-19 pandemic should attempt to make ultrasound available for cardiac assessment, which would improve RHD diagnosis and potentially strengthen risk assessment for those presenting with suspected COVID-19 infection. Further, in countries where RHD is endemic, there may be some diagnostic overlap between COVID-19 infection and new or recurrent acute respiratory failure, which can present with fever, joint pain, and sore throat, though typically lacking prominent respiratory distress and hypoxia associated with COVID-19 infection.

### The indirect risk of COVID-19 infection to people living with RHD

In contrast to the direct risk of COVID-19 to people living with RHD, which is largely unknown, there are known indirect risks of the COVID-19 pandemic to people living with RHD, largely driven by the global disruption in healthcare systems and supply chains. RHD patients are sensitive to receiving regular and timely care—and disruptions to service access increases risk of progression and adverse outcomes.

Secondary prophylaxis with continuous antibiotic coverage to prevent recurrent group A streptococcal (GAS) infection, recurrent rheumatic fever, and worsening RHD is the cornerstone of guideline-based management for RHD. Benzathine penicillin G (BPG), delivered through regular intramuscular injection, typically every 28 days, provides the best protection and is standard of care for those without penicillin allergy.

Receipt of secondary prophylaxis requires frequent interaction with the healthcare system. Globally, transportation restrictions have limited public and private vehicles, greatly reducing healthcare access. Governments should recognize the need for secondary prophylaxis and ensure that patients with RHD receive medical clearance/approval to be on the road. Further, local staffing constraints at healthcare facilities may present an additional challenge to receipt of secondary prophylaxis. As soon as possible, health authorities should prioritize RHD secondary prophylaxis clinics as essential care and strive to have no interruption of prophylaxis delivery. It would be reasonable, given presumed increased risk of COVID-19 to patients living with RHD, to try to ensure that physical distancing is maintained with all RHD patients. It will also be important to monitor the global and local BPG supply chains. Qualitative data suggest no major supply chain disruptions have yet occurred, but all of the active pharmaceutical ingredient (API) for penicillin formulations is manufactured in China, and the global supply chain, even prior to COVID-19, was considered fragile.

Though not considered optimal, oral penicillin prophylaxis should be considered for patients who are unable to reliably receive intramuscular BPG. While unequivocally inferior, oral penicillin would provide reasonable protection against recurrent GAS infection and would be

**Table 1** Summary of recommendations for patients, providers, and health systems dealing with RHD in the time of the COVID-19 pandemic

1. Patients living with RHD should, to the best of their ability, follow the basic infection control recommendations provided by the World Health Organization as well as their local and national governments.
2. Patients living with RHD should continue to take all prescribed cardiac medications, including those classified as angiotensin-converting enzyme inhibitors (ACEIs) and angiotensin II receptor blockers (ARBs).
3. Governments should recognize the need for secondary prophylaxis and ensure that patients with RHD receive medical clearance/approval to be on the road.
4. Health authorities should prioritize RHD secondary prophylaxis clinics as essential care and strive to have no interruption of prophylaxis delivery.
5. Providers should consider giving oral penicillin prophylaxis as a temporary solution for patients who are unable to reliably receive intramuscular BPG.
6. Providers should put in place a care plan for patients whose surgery has been delayed due to COVID-19 constraints including developing robust systems for prioritization.

far superior to no antibiotic prophylaxis. Health systems and providers who are considering moving patients from intramuscular to oral prophylaxis should see this measure as temporary and emphasize to patients that return to intramuscular prophylaxis provides superior protection and should be resumed as soon as possible.

The delay in surgical and catheter-based intervention for patients living with RHD will almost certainly cause indirect harm. In-country 'elective' surgery has largely stopped in many places, and travel bans will limit the number of patients who can receive care abroad, or receive visiting teams, mainstays for many LMICs. Health systems will need to adapt, including developing robust systems for prioritization. Ultimately strengthening diagnostic access for RHD, such as programmes to employ decentralized echocardiography, could provide a better solution, allowing people living with RHD to be identified at earlier stages, when secondary prevention may prevent progression to surgical disease.

Finally, the COVID-19 pandemic threatens progress in RHD research, advocacy, and country programmes. In 2018, the World Health Assembly passed a global RHD resolution, reprioritizing RHD on the global agenda. Countries working to integrate National Action Plans, will face renewed challenges for implementation given the resource diversion to COVID-19. While this diversion is appropriate, past global surges in RHD control (such as the WHO activities of the 1980s) were eroded with other disease control priorities, and the RHD and global community will need to be mindful so that once

COVID-19 is controlled, we do not lose the recent gains that have been made to prioritize RHD (Table 1).

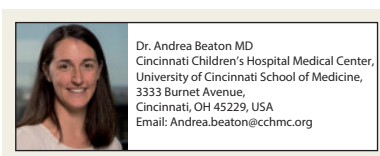
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## References

References are available as [supplementary material](#) at *European Heart Journal* online.



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# PEARS procedure and the difficulty to provide evidence for its benefits

**Personalised External Aortic Root Support (PEARS) in patients with a dilated aortic root and ascending aorta was established some time ago although is still not fully recognised by the cardiovascular community.<sup>1</sup>**

The ExoVasc<sup>®</sup> Personalised External Aortic Root Support (PEARS) is a computer designed, custom-made implant to match individual aortic root morphology. It was first introduced in 2004 as a conservative surgical approach for Marfan patients with asymptomatic aortic dilation.<sup>2-4</sup> The concept and production of a personalized device was the

result of research and development between 2000 and 2004 when the first operation was performed.<sup>2,5</sup>

Computer-aided design based on three-dimensional re-construction of magnetic resonance/computer tomographic (MR/CT) images, a dedicated manufacturing method, and a refined surgical technique