# Rheumatic heart disease

## *Executive summary*

## Introduction

Rheumatic heart disease (RHD) is a serious complication of rheumatic fever. It refers to the chronic heart valve damage that can develop years after single or repeated episodes of acute rheumatic fever (ARF). Acute rheumatic fever is an inflammatory autoimmune disease which occurs following inadequately treated infection with Group A Streptococcus (GAS). Repeated episodes of ARF can lead to inflammation of the heart either on the surface of the heart (pericarditis), within the heart (endocarditis) leading to accumulative damage to one or more of the four heart valves (endocarditis) or involving the heart muscle itself (myocarditis). Untreated RHD can lead to heart failure, stroke, arrhythmia and complications in pregnancy.

RHD is a disease of poverty, associated with overcrowding and poor living conditions, as well as lack of access to health care and services.

Group A streptococcal infection, acute rheumatic fever and rheumatic heart disease are preventable with improved housing and living conditions, and early antibiotic treatment.

## Target users

* Doctors
* Nurses

## Target area of use

* Outpatient department
* Ward

## Key areas of focus / New additions / Changes

This guidelines describes the initial diagnosis and management of rheumatic heart disease. Advanced rheumatic heart disease requires surgical treatment which is delivered through a partnership with the UK charity Chain of Hope. Management of these children is shared with overseas paediatric cardiologists who will advise on more complex problems.

## Limitations

Access to surgery is limited and many children must be managed medically.

## Presenting symptoms and signs

The most common forms of RHD include mitral valve disease (mitral stenosis, mitral regurgitation), aortic valve disease (aortic stenosis, aortic regurgitation) and tricuspid regurgitation. Atrial fibrillation is often associated with mitral stenosis.

Clinical features of RHD vary depending on which heart valves are affected and how severely. Early in the onset of disease, cases may be asymptomatic for many years. As valvular disease progresses, cases will have symptoms of some degree of heart failure, ranging from mild to severe exertional dyspnoea, fatigue, peripheral oedema and palpitations. The severity of RHD increases with repeated episodes of ARF.

Complications of RHD include heart failure, stroke, arrhythmias, infective endocarditis and complications during pregnancy. Heart failure is the major cause of disability and death from RHD. Infective endocarditis is more likely to occur when there is damage to the heart valves and is a serious illness requiring hospitalisation and intravenous antibiotics, which is also associated with a significant mortality and morbidity burden.

Other symptoms due to rheumatic heart disease are:

* Chest pain.
* Heart palpitations.
* Shortness of breath especially with physical exertion.
* Swollen ankles, legs, wrists, and stomach.
* Excessive fatigue.
* Thumping sensation in chest.

## Examination findings

These vary with the severity of valve calcification, the severity of stenosis, and left ventricular (LV) function.

With mild disease, the apex beat is brisk and hyper dynamic. The S1 is usually soft, and a widely split S2 is common. A diastolic rumble and S3 may be present even if there is no LV dysfunction.

With severe mitral regurgitation (MR), the apex may be enlarged and displaced laterally. The systolic murmur of MR varies according to its cause. It is usually heard best at the apex in the left lateral decubitus position. With severe degenerative MR, the murmur is holosystolic, radiating into the axilla. Early systolic murmurs are typical of acute MR. Late systolic murmurs are typical of mitral valve prolapse or papillary muscle dysfunction.

Signs of pulmonary hypertension, such as a loud P2, represent advanced disease.

## Differential diagnosis

The main alternative diagnosis in children is infectious endocarditis or congenital heart disease. In adults, valvular heart disease might be due to degenerative changes.

## Investigations

* **CXR** 
  + Heart failure: enlarged heart, congested pulmonary vasculature. If more severe: enlarged right atrium and right ventricle.
  + Chronic MR: cardiomegaly (LV and LA enlargement).
  + Acute MR or progressive LV failure: Kerley B lines and interstitial oedema.
  + Pulmonary hypertension: right-sided chamber enlargement.
  + Severe mitral stenosis: left atrial enlargement.
* **ECG** – Looking for LV hypertrophy, LA enlargement, conduction abnormality (left and right bundle branch block), altered axis, atrial fibrillation.
* **ECHO** – Essential to confirm the diagnosis and grade the severity of disease.

Other investigations are not available in the Gambia.

## Management

There are 2 goals in treating RHD:

1. Prevention of further damage to the heart valves.
2. Treatment of heart failure.

### Prevention of further damage to the valves

Primary prevention involves the use of antibiotics to treat GAS throat infections.

Secondary prevention of ARF: once a person has had ARF, it is important to prevent future episodes, this is best provided by monthly IM injections of Benzathine penicillin at a dose of 1.2 MU. This is increased to 3 weekly if the patient is at especially high risk. It should be continued until 10 years after the last episode of ARF or the age of 21 years. This treatment reduces the number of ARF episodes and should be offered to all patients with rheumatic carditis. For persons with established rheumatic heart disease, secondary prevention with continuous antibiotics should be continued for longer periods – even for life. It is also continued even after valve surgery.

Penicillin V has been used in place of Benzathine penicillin. Patients prefer it, but there is concern that it is less effective because it depends on them taking medication every day. There is no evidence to support its use.

### Treatment of heart failure

Medical treatment

**ACE inhibitors** block the formation of angiotensin II, this medication lowers blood pressure and reduces sodium retention in people who have heart failure. They are relatively contraindicated in patients with mitral or aortic stenosis.

Use enalapril 100 micrograms/kg OD initially. The dose can be increased up to 1 mg/kg OD. The maximum dose is 10 mg OD. Ramipril or captopril can be substituted according to availability.

**Beta blockers** block the effect of adrenaline on the heart to reduce excess stress on the failing heart. These medications are most commonly used for dilated cardiomyopathy.

Use bisoprolol 1.25 mg OD in older children. Use in younger children only on the advice of a cardiologist.

**Diuretics** stimulate the body to lose water by acting on the kidneys. This effect decreases fluid retention and edema and provides symptomatic relief to the patient.

Start with furosemide 0.5 mg/kg OD. The dose can be increased as high as 12 mg/kg, but it is unusual to need doses higher that 2 mg/kg OD.

Add spironolactone 1 mg/kg OD. The dose can be increased up to 9 mg/kg OD, but usually lower doses are needed in heart failure.

Surgical treatment

Patients with significant heart failure should be considered for valve repair or replacement surgery. In an ideal world, even asymptomatic patients should be considered for this in order to prevent significant heart damage.

Patients are often very young when operated and, therefore, are given mechanical valves if they need replacement surgery, as they last longer. However, they then require warfarin treatment. Girls offered mechanical valves should be counselled about the risks of being pregnant on warfarin treatment and about the option of using low-molecular weight heparin through the pregnancy. The target INR is usually 2.5-3.5.

## Key Issues for Nursing care

## Make sure patients and their caregivers understand why they have been given penicillin prophylaxis and how important it is to prevent further episodes of ARF.

## References

Queensland Health Guidelines

Maganti, K., Rigolin,V.H., Sarano, M.E.,MD, and Bonow, R.O. Valvular Heart Disease: Diagnosis and Management. Mayo Clin Proc. 2010. 85(5): 483–500.

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