Walking the tightrope: when to intervene in Rheumatic heart disease

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Learning objectives

- To systematically evaluate the severity of valvular pathology in rheumatic heart disease (RHD) and its impact on left ventricular (LV) function.
- To ascertain when surgical intervention is needed to preserve ventricular function.
Background

Rheumatic fever is a disease of poverty. Poor socio-economic circumstances including overcrowding, malnutrition and limited access to primary health care contribute to this epidemic in sub-Saharan Africa and other third world economies [1].

Children aged 5-14 years develop acute rheumatic fever after upper respiratory infection with Group A #-haemolytic streptococcus [2]. These organisms attach to the upper respiratory tract epithelial cells and produce enzymes which damage and invade epithelial cells eliciting an inflammatory response. The M surface protein is a virulence antigen structurally similar to cardiac muscle tissue myosin and cardiac valve laminin. Anti-M antibodies formed against streptococcal antigens cross-react with these tissues resulting in myocarditis, pericarditis and endocarditis with valvular lesions [3].

The clinical diagnosis of RHD is based on the Modified Jones criteria and more recently the World Health Organization (WHO) criteria [2].

Mitral regurgitation (MR) and aortic regurgitation (AR) are the two most common valvular lesions in RHD affecting young patients between the ages of 6 -18 at our institution. MR often occurs with mitral stenosis (MS), and similarly with AR and aortic stenosis (AS). Progressive fibrosis and partial fusion of the valve leaflets occur, thus MS and AS often present in the third decade of life [3].

In our series, children as young as six years have established, severe RHD necessitating prosthetic valve replacement. However, due to their small annular size, placing a prosthesis that will be adequate for future growth is impossible and serial valve replacements are needed. This contributes significantly to their morbidity and mortality. Delaying surgery until the child is older is also not advisable as the progressive dilation of the left ventricle inevitably leads to its dysfunction. Vigilant monitoring of the valvular pathology and left ventricle (LV) size and function is thus needed for timeous surgical intervention.
Imaging findings OR Procedure details

Chest radiography is usually the first radiological investigation where impressions of great vessels, cardiac size, chambers, appendages as well as airway and lungs can be derived.

Enlarged left atrium (LA) affects the cardiac silhouette and adjacent structures. Left atrial hypertrophy (LAH) appears as 'double density' on frontal chest radiograph (fig. 1) and posterior protrusion of the left atrial border on lateral chest radiograph (fig. 2). The increased carina angle can be appreciated on frontal chest radiograph. On frontal chest radiograph, enlarged left ventricle (LV) may displace the apex of the heart to the left and inferiorly (fig. 1) whereas on the lateral view, the inferior posterior cardiac border is displaced to meet the IVC line below the diaphragm (fig. 2). Right ventricle enlargement is best appreciated in the lateral view where filling of the retrosternal space can be demonstrated (fig. 2). Calcification of the valves and pulmonary congestion can also be visualized on chest radiograph [6].

Recent data suggests that echocardiography should be routinely used as a screening and diagnostic tool for borderline and definitive RHD. In 2007, a study done in Mozambique and Cambodia by Marijon et al concluded that the use of echocardiography in diagnosing RHD is superior to that of clinical examination alone. Approximately 90% of clinically silent cases were identified as positive on echocardiography [4]. More recent study conducted by Shah et al in India revealed similar results [5]. Dr Liesl Zuhlke from Red Cross Children’s Hospital in SA is currently busy with a similar study.

The World Heart Federation (WHF) recently published evidence-based criteria for echocardiographic diagnosis of subclinical and clinical RHD as described in table 1 and 2 [3].

<table>
<thead>
<tr>
<th>MR</th>
<th>MR</th>
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| Doppler | • Seen in two views  
• At least one view, jet length $\geq$ 2 cm  
• Velocity $\geq$ 3 m/s for one complete envelope  
• Pan-systolic jet in at least one envelope |
| Morphological features | • Anterior mitral valve leaflet (AMVL) thickening $\geq$ 3 mm for $\leq$ 20 years |
• Chordal thickening
• Restricted leaflet motion
• Excessive leaflet tip motion during systole

Table 1: WHF Echocardiographic criteria for pathological mitral regurgitation (MR) [3]

<table>
<thead>
<tr>
<th>Doppler</th>
<th>AR</th>
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<tbody>
<tr>
<td>• Seen in two views</td>
<td></td>
</tr>
<tr>
<td>• At least one view, jet length ≥ 1 cm</td>
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<tr>
<td>• Velocity ≥ 3 m/s in early diastole</td>
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<tr>
<td>• Pan-diastolic jet in at least one envelope</td>
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<tr>
<th>Morphological features</th>
<th>AR</th>
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<tr>
<td>• Irregular or focal thickening</td>
<td></td>
</tr>
<tr>
<td>• Coaptation defect</td>
<td></td>
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<tr>
<td>• Restricted leaflet motion</td>
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<td>• Prolapse</td>
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Table 2: WHF Echocardiographic criteria for pathological AR [3]

At our institution, we correlate body mass index (BMI) of the patient to M-mode echocardiographic measurements when screening and diagnosing left heart enlargement due to RHD. Z-scores of the normal LV measurements on M-mode for children are also widely available making it possible to determine the degree of deviation from the average for the age and size of the patient. Accurate LV measurement on M-mode must be obtained to determine timing for surgical intervention [3].

The decision of when to intervene surgically in a child with RHD is based on the clinical picture and findings on echocardiography. As demonstrated in the research above, clinical examination may under-diagnose valvular pathology and thus many of our patients are only referred when they have advanced RHD with cardiac failure. Serial echocardiographic assessment of patients at risk of developing RHD should be performed 3 - 6 monthly [3]. 2D morphology of the mitral and aortic valves, LV measurements and function on M-mode (fig. 3) and colour Doppler interrogation of the involved valves should be routinely evaluated [3]. 3D echocardiography is increasingly being used in surgical centres to evaluate the morphology of the affected valve and its suitability for surgical intervention [3].

Patients with severe cardiomegaly and/ or depressed LV function have an increased surgical risk, are less likely to regain normal cardiac function after surgery and have an
increased risk of late cardiac failure and death. It is thus recommended that patients, especially children, should be referred for surgery once the parameters outlined in table 3 are approached, regardless of their clinical symptoms [3].

<table>
<thead>
<tr>
<th>Clinical criteria</th>
<th>Severe MR</th>
<th>Severe AR</th>
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<tr>
<td></td>
<td>• New York Heart Association Functional Capacity (NYHA FC) II-IV</td>
<td>• NYHA FC II-IV</td>
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<tr>
<td></td>
<td>• Cardiomegaly</td>
<td>• Widened pulse pressure &gt; 50 mmHg OR &gt; 50% of systolic pressure</td>
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<td>• Pulmonary hypertension</td>
<td>• Signs of severe AR: De Musset, Quincke signs or pistol shots audible over arteries</td>
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<td>2D criteria</td>
<td>• Thickening of mitral leaflets, chordae and papillary muscles (fig. 4)</td>
<td>• Thickening and retraction of aortic valve leaflets</td>
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<td>• Shortening and tethering of posterior mitral valve leaflet (PMVL) (fig. 4)</td>
<td>• Lack of normal coaptation of leaflets</td>
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<tr>
<td></td>
<td>• Anterior mitral valve leaflet (AMVL) may be thickened and retracted or elongated and prolapsing past PMVL</td>
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<td></td>
<td>• Excessive leaflet tip motion during systole in keeping with chordal rupture (fig. 4)</td>
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<td>• Lack of normal coaptation of leaflets (fig. 5)</td>
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### M-mode criteria
- Left ventricular ejection fraction (LVEF) < 60%
- Left ventricular inner diameter in systole (LVIDs) > 35 mm OR > +2 Z-score for BMI or weight

### Colour Doppler criteria
- Pan-systolic MR jet seen in two views
- Parasternal long axis view: Posteriorly-directed MR jet is characteristic of RHD (fig. 6)
- 4 chamber view: Jet length ≥ 2/3 of LA length; MR jet may extend back into dilated pulmonary veins (Fig. 7)
- Broad AR jet seen in two views
- Parasternal long axis view: Posteriorly-directed AR jet along AMVL is characteristic of RHD (fig. 8)
- 4 chamber view: Jet length ≥ 2/3 of LV cavity length (fig. 9)
- Suprasternal view: diastolic flow reversal in descending aorta

### Doppler flow criteria
- Pulmonary hypertension with estimated PA systolic pressure ≥ 40 mmHg
- Pan-diastolic envelope of AR
- Velocity time index (VTI) % reversed flow in descending aorta > 50% (retrograde flow VTI/anterograde flow VTI 100)

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<th>Table 3: Indications for surgical intervention in MR and AR [2]</th>
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<td>Patients with less severe RHD can undergo valvular repair. However, patients with severe, established fibrosis and destruction of the valves can only be offered valvular replacement surgery and will thus need lifelong anticoagulation therapy [3].</td>
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Fig. 1: Frontal chest radiograph demonstrating LAH, LVH and pulmonary plethora.

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Fig. 2: Lateral chest radiograph demonstrating LVH and RVH.

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**Fig. 3:** M mode long axis echocardiograph demonstrating LV enlargement and function.

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**Fig. 4:** 2D 4 Chamber echocardiograph view demonstrating MV leaflet and chordal thickening with lack of coaptation: AMVL prolapses past shortened, tethered PMVL.
Fig. 5: 3D echocardiograph demonstrating lack of MV coaptation.
Fig. 6: Colour Doppler 2D long axis echocardiograph view demonstrating posteriorly directed MR jet.

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Fig. 7: Colour Doppler 2D 4 chamber echocardiograph view demonstrating posteriorly directed MR jet.

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Fig. 8: Colour Doppler 2D long axis echocardiograph view demonstrating posteriorly directed AR jet impinging on AMVL.

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**Fig. 9:** Colour Doppler 2D 4 chamber echocardiograph view demonstrating posteriorly directed AR jet impinges on AMVL and MR jet to dilated LA.

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Conclusion

RHD is a major cause of morbidity and mortality affecting nearly 20 million patients worldwide in developing countries such as South Africa [7]. Prevention and early recognition is the key to reducing the high prevalence of this disease in our developing nations. Current research highlights the importance and sensitivity of echocardiography in diagnosing the presence and severity of RHD. Serial echocardiographic follow up is essential to monitor for progression of disease and to evaluate when referral for intervention is indicated.
References