Any person with suspected ARF and a cardiac murmur, or any case of chorea, should have an echocardiogram shortly after admission to hospital.

Echocardiogram:
- Normal
- Repeat at 2-4 weeks
- Abnormal

Notes 1 & 2
- Pursue alternative diagnoses

Notes 3 & 4
- Second echocardiogram at 2-4 weeks if no other alternative diagnosis. A second echo is usually unnecessary with a presentation of chorea.

Echocardiogram:
- Normal
- Repeat at 2-4 weeks
- Abnormal

Notes 1 & 2
- Second echocardiogram at 4-6 weeks if:
  - Signs progress
  - Medication commenced
  - Recommended by cardiologist
Note 1
Minimal Echocardiographic Criteria to Allow a Diagnosis of Pathological Valvular Regurgitation

AORTIC REGURGITATION

- Colour: Substantial colour jet seen in 2 planes extending greater than or equal to 1 cm beyond the valve leaflets
- Continuous wave or pulsed Doppler: Holosystolic with well-defined high velocity spectral envelope

MITRAL REGURGITATION

- Colour: Substantial colour jet seen in 2 planes extending greater than or equal to 2 cm beyond the valve leaflets
- Continuous wave or pulsed Doppler: Holosystolic with well-defined high velocity spectral envelope

If the aetiology of aortic or mitral regurgitation on Doppler echocardiography is not clear, the following features support a diagnosis of rheumatic valve damage:

- Both mitral and aortic valves have pathological regurgitation
- The mitral regurgitant jet is directed posteriorly, as anterior mitral valve prolapse is more common than posterior valve prolapse
- Multiple jets of mitral regurgitation
- The presence of morphological or anatomical changes consistent with RHD (see guideline), but excluding slight thickening of valve leaflets:
  - definite thickening of mitral valve leaflets, indicative of chronic RHD
  - elbow or dog leg deformity of anterior mitral valve leaflets

Echocardiography allows the operator to comment on the appearance of valves that are affected by rheumatic inflammation. The degree of thickening gives some insight into the duration of valvulitis, no significant thickening occurring in the first weeks of acute rheumatic carditis (Level IV evidence - see guideline)

**
Only after several months is immobility of the subchordal apparatus and posterior leaflet observed. Several other findings have also been reported, including acute nodules, seen as a beaded appearance of the mitral valve leaflets. Although none of these morphological features are unique to ARF, the experienced echocardiographic operator can use their presence as supportive evidence of a rheumatic aetiology of valvulitis.

Source: Adapted with permission from Wilson, N.J. & Neutze, J.M.

Note 2
Severity of ARF Carditis

MILD CARDITIS*

- Mild mitral or aortic regurgitation clinically and/or on echo (fulfilling the minimal echo standards in Note 1) with no clinical evidence of heart failure and no evidence of cardiac chamber enlargement on CXR, ECG or echocardiography

MODERATE CARDITIS

- Any valve lesion of moderate severity clinically (e.g. mild or moderate cardiomegaly), or
- Any echocardiographic evidence of cardiac chamber enlargement or any moderate severity valve lesion on echo**:• Mitral regurgitation is considered moderate if there is a broad high-intensity proximal jet filling half the left atrium or a lesser volume high-intensity jet producing prominent blunting of pulmonary venous inflow
• Aortic regurgitation is considered moderate if the diameter of the regurgitant jet is 15% to 30% of the diameter of the left ventricular outflow tract with flow reversal in upper descending aorta

SEVERE

- Any impending or previous cardiac surgery for RHD, or
- Any severe valve lesion clinically (significant cardiomegaly expected, and/or heart failure), or
- Any severe valve lesion on echo:
  - Abnormal regurgitant colour and Doppler flow patterns in pulmonary veins are a prerequisite for severe mitral regurgitation
  - Doppler reversal in lower descending aorta is required for severe aortic regurgitation

Note 3
Differential Diagnoses of Common Major Manifestations of ARF

<table>
<thead>
<tr>
<th>POLYARThRitis AND FEVER</th>
<th>CARDITIS</th>
<th>PRESENTATION</th>
<th>CHOREA</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Differential diagnoses</strong></td>
<td><strong>Other infections</strong> (incl. gonococcal)</td>
<td><strong>Innocent murmur</strong></td>
<td><strong>Systemic lupus erythematosus</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Connective tissue and other auto-immune disease</strong> **</td>
<td>** <strong>Mitral valve prolapse</strong></td>
<td><strong>Drug ingestion (extrapyramidal syndrome)</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Reactive arthropathy</strong></td>
<td><strong>Congenital heart disease</strong></td>
<td><strong>Wilson’s disease (usually adult onset)</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Sickle cell anaemia</strong></td>
<td>** Infective endocarditis**</td>
<td><strong>Tic disorder (see guideline)</strong></td>
</tr>
<tr>
<td></td>
<td>** Infective endocarditis**</td>
<td>** Hypertrophic cardiomyopathy**</td>
<td><strong>Congestional, e.g. hyperbilirubinaemia</strong></td>
</tr>
<tr>
<td></td>
<td>** Leukaemia or lymphoma**</td>
<td><strong>Myocarditis — viral or idiopathic</strong></td>
<td><strong>Choreaathetoid cerebral palsy</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Gout and pseudogout</strong></td>
<td><strong>Pericarditis — viral or idiopathic</strong></td>
<td><strong>Encephalitis</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Henoch-Schonlein purpura</strong></td>
<td></td>
<td><strong>Familial chorea (including Huntington’s)</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Post-streptococcal reactive arthritis</strong> **</td>
<td>**</td>
<td><strong>Intracranial tumour</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Other, e.g. HIV/AIDS, leukaemia</strong></td>
<td></td>
<td><strong>Hormonal</strong></td>
</tr>
</tbody>
</table>

Notes:

* Includes bacterial arthritis, influenza B, cytomegalovirus, Epstein-Barr Virus, mycoplasma, rubella (also post-vaccination), hepatitis B, parvovirus, Vzv are all others except those with gastrointestinal histopathological

** Includes rheumatoid arthritis, juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis and sarcoidosis, among others

*** In these cases the arthritis may affect joints that are not commonly affected in ARF (such as the small joints of the hand), and is less responsive to anti-inflammatory treatment. It is recommended that the diagnosis of post-streptococcal reactive arthritis should rarely, if ever, be made in high-risk populations and with caution in low-risk populations (Grade C evidence - see guideline)

# Drugs and toxic: include anticonvulsants, antidepressants, lithium, scopolamine, calcium channel blockers, methylphenidate, theophylline and antihistamines

§ Includes oral contraceptives, pregnancy (chorea gravidarum), hyperthyroidism and hyperparathyroidism.