Acute Rheumatic Fever

Patient Teaching Tool

Jodi Mann, Rhona Dawson and KAMS Healthy Communities Team
Acute Rheumatic Fever (ARF) is a delayed autoimmune reaction following an untreated Group A Streptococcal (GAS) infection of the upper respiratory tract/throat and sometimes from skin sores.

It predominantly affects children aged 5-14 years.

The immune mediated damage is an acute generalised inflammatory response, affecting certain parts of the body - the heart, joints, brain and skin.

People who have had ARF previously are more likely to have further episodes of ARF (recurrences).

Not everyone is susceptible to developing ARF and not all GAS strains are "rheumatogenic". But antibiotic treatment of GAS infections (throat/skin) in high risk populations like Aboriginal and Torres Straight Islander people is essential (Primary Prevention of ARF).

There is currently no vaccine available to treat GAS - but research is underway.
What is Acute Rheumatic Fever (ARF)?

It is a sickness caused by a germ called ‘Streptococcus’ generally known as [STREP].

The [STREP] germ is a common cause of sore throats and skin sores in children aged 5-14 yrs.

In some children [STREP] throat and skin sores can cause sickness in other parts of the body - joints, heart, skin and brain.

This is known as Acute Rheumatic Fever - [ARF].
DIAGNOSIS OF ARF – MODIFIED JONES CRITERIA

No specific laboratory diagnostic test. Diagnosis based on clinical criteria.

- Signs and Symptoms of ARF:
  - fever $\geq 38$.
  - joint pain (+ or- swelling)- usually large joints especially knees and ankles (can be asymmetrical and migratory).
  - jerky uncontrolled movements (Sydenham's chorea) which can be mild to severe especially of the hands, feet, tongue and face.
  - carditis- damage to the heart valves usually the mitral (most common) and/or aortic heart valve- determined by echocardiogram (a heart murmur may only be present with significant valve damage).
  - subcutaneous nodules and rash (Erythema marginatum) are rare 2% of all cases.

- Joint pain and/or Sydenham's Chorea can be common presentations of ARF.
- All joint, skin and brain manifestations (Sydenhams Chorea) will get better with time.

**DAMAGE TO THE HEART VALVES MAY BE PERMANENT.**

- Damage to the heart valves can occur during the initial episode of ARF.
- Recurrent episodes of ARF can cause significant heart valve damage and compromise.
Acute Rheumatic fever (ARF) is a sickness caused by a germ called *STREP*. It can affect the heart, joints, skin and brain.
Clinical Tests required for diagnosis of ARF at time of presentation:

- Throat or skin swab/slide- MC&S (GAS)
- Temperature
- ECG
- Bloods
  - Full blood count (FBC)
  - Erythrocyte Sedimentation Rate (ESR)
  - C-Reactive Protein (CRP)
  - Anti-streptococcal Serology ie Antistreptolysin (ASOT) and anti-DNase (antiDNase) titres
    Repeat 10-14 days later if first test not confirmatory.
- Echocardiogram and review by a cardiologist at the time of presentation or within 3 months.
Testing for ARF:
ARF is caused by an untreated group A streptococcus (GAS) infection of the upper respiratory tract (URT).

There is a latent time period on average of 3 weeks from the time of infection with GAS to the time when symptoms of ARF develop.

At the time of an untreated GAS infection antibodies produced by the human immune system (bodies defense army to kill germs) is unable to distinguish between the bodies self proteins and the invading GAS antigen (germ).

This triggers an inflammatory autoimmune response which attacks self proteins of the joints, heart, brain and skin = ARF.
How **ARF** affects the body:

Your body produces good germs (antibody) to attack the **STREP** germs (antigen).

The **STREP** germs confuse the good germs.

The good germs then attack your heart valves, joints, skin and brain - thinking they are attacking **STREP**.

Most of the symptoms like fever, sore and swollen joints, skin rash and jerky movements go away with time - but damage to your heart may be permanent.
■ Damage to the heart caused by Acute Rheumatic Fever (ARF) is known as Rheumatic Heart Disease (RHD).

■ Unlike all the other manifestations of ARF, RHD may be permanent and cause significant cardiac (heart) damage.

■ RHD can present on the initial episode of ARF.

■ Recurrent episodes of ARF significantly increase the severity of RHD.

■ A person can have ARF without having RHD, but cannot have RHD without having had ARF.
The damage to your heart caused by ARF is known as Rheumatic Heart Disease (RHD).
Rheumatic Heart Disease (RHD) is the result of ARF and can cause significant illness and death.

RHD affects the mitral valve (most common) and/or aortic valve of the heart (identify on pictures). Tricuspid valve disease is uncommon and almost always associated with mitral valve disease.

The heart valve is like a “door” opening and closing, assisting the blood being pumped by the heart to move in one direction. Damage to these “doors” (heart valves) lets the blood flow back in the wrong direction (valve regurgitation) or restrict the flow forwards through the heart valve (valve stenosis).

If the valve is severely affected with RHD, open heart surgery may be required to either replace or repair the valve.

The main goal for management of RHD is the prevention of further episodes of ARF = via secondary prophylaxis (Long Acting Benzyl Penicillin (Bicillin LA)- Intra muscular (IM) injection every 21-28 days).
Superior vena cava
Inferior vena cava
Pulmonary veins
Aorta
Right ventricle
Left ventricle
Mitral valve
Aortic valve
Right atrium
Left atrium
Pulmonary artery
Pulmonary veins
Tricuspid valve
Pulmonary valve
Aortic valve
Left ventricle
Right ventricle
Inferior vena cava

Normal Heart

Damaged Heart

Damaged valves

RHD
We cannot stop the autoimmune response the body produces (ARF) but we can stop the body being infected by the GAS germ that causes the response = we do this with the Long Acting Benzyl Penicillin (Bicillin LA) known as Secondary Prophylaxis every 21-28 days.

After 28 days the person is not protected against a GAS infection and the development of ARF.

Long Acting Benzyl Penicillin (Bicillin LA)- Intra muscular (IM) injection every 21-28 days.

**WHY?**
- prevent further GAS infections
- prevent recurrences of ARF
- to prevent the development or worsening of RHD
- reduce the severity of RHD
- reduce the risk of death from RHD.

**DOSAGE**

<table>
<thead>
<tr>
<th>Weight</th>
<th>Dosage</th>
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</thead>
<tbody>
<tr>
<td>Less than 20kg</td>
<td>450mg of Bicillin LA</td>
</tr>
<tr>
<td>20 kg and more</td>
<td>900mg of Bicillin LA</td>
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</tbody>
</table>

**Length of Secondary Prophylaxis**

<table>
<thead>
<tr>
<th>Priority</th>
<th>Duration</th>
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<tbody>
<tr>
<td>Priority 3 mild/no RHD</td>
<td>Continue until the age of 21 or 10 years since last episode of ARF (which ever is longer).</td>
</tr>
<tr>
<td>Priority 2 moderate RHD</td>
<td>Until the age of 35 years.</td>
</tr>
<tr>
<td>Priority 1 severe RHD</td>
<td>Until the age of 40 years or longer (life).</td>
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The decision regarding the duration of secondary prophylaxis is based on many factors including echocardiogram and is best determined by a Cardiologist/Physician.
How do we stop damage to your heart?

Stop from getting ARF again.

How do we do this?

By having a needle/ Bicillin every 21-28 days.
Almost all cases of ARF and associated RHD are completely preventable

Treat and prevent GAS infections:

- keep yourself clean
- wash your face and hands regularly
- eat healthy foods
- brush your teeth every day
- keep your community clean.

BUT MOST IMPORTANTLY:

- go to see the nurse or doctor for sore throats and skin sores (all members of the family).
- always take all your medicine given to you by your nurse or doctor.
What else can you do?
Secondary prophylaxis adherence

<table>
<thead>
<tr>
<th>Factors that influence poor adherence to secondary prophylaxis:</th>
<th>Factors that influence good adherence to secondary prophylaxis:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• needles are painful!</td>
<td>• injections given slowly and correctly</td>
</tr>
<tr>
<td>• lack of education about importance/disease</td>
<td>• active recall before 28 days</td>
</tr>
<tr>
<td>• embarrassment/ shame</td>
<td>• establishing a clinic/ health professional rapport with the</td>
</tr>
<tr>
<td>• reluctance to wait or attend a health facility</td>
<td>patient</td>
</tr>
<tr>
<td>• other priorities in life</td>
<td>• ongoing education about importance of needles and ARF/</td>
</tr>
<tr>
<td>• unable to calculate 28 days</td>
<td>RHD.</td>
</tr>
<tr>
<td>• don’t feel sick.</td>
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</tbody>
</table>

**Bacterial Endocarditis Prophylaxis**

Bacterial Endocarditis is a dangerous complication of RHD. Anyone with a prosthetic valve or established RHD should receive additional antibiotic prophylaxis prior to procedures expected to produce bacteremia - dental, oral and some respiratory tract procedures.
Important

It is important to NEVER miss a needle / Bicillin.

- If you do, have it as soon as you can.
- Tell your nurse, doctor or health worker if you are moving house or town.
- Attend all your doctors’ appointments.
- Tell the dentist that you have Acute Rheumatic Fever / Rheumatic Heart Disease.
FOR MORE INFORMATION
Refer to “The Australian guideline for the prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease” (2nd edition) for further information.

Available from:
www.rhdaustralia.org.au

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