

CHIA MEDICAL TEAM RESOURCE PROTOCOLS:

Rheumatic Heart Disease

It is not very common for children to get an acquired heart disease (meaning that they weren't born with it). In adults, acquired heart disease is caused by lifestyle factors, such as obesity, high blood pressure, and high cholesterol that damage the heart and cause heart disease.

However, children can get acquired heart disease when their heart is damaged by inflammation caused by an infection called Acute Rheumatic Fever. This is called Rheumatic Heart Disease (RHD).

Acute Rheumatic Fever:

Acute Rheumatic Fever is caused by bacteria called Group A Streptococcus that typically infect the throat. Not all strains of streptococcus bacteria will cause serious illness. In some people, infection with the Streptococcus bacteria causes a mild disease, commonly called "step throat". Step Throat very common in children and is normally very mild. Symptoms include fever (>38 degrees), sore throat and enlarged tonsils with pus on them. For most people it causes no complications, however it should be treated with antibiotics. In some people, however, infection with strep throat can cause an immune response that results in Acute Rheumatic Fever (ARF). It can be several weeks after an initial strep throat infection before symptoms of ARF begin. Scientists don't really understand why some people who get strep throat also get ARF. About 3-5% of people are born with extra susceptibility to getting ARF. ARF is most common in children aged 5 to 15. ARF causes inflammation in the heart, joints, skin or central nervous system. Children who get ARF are often very unwell, and require hospital treatment.



Figure 1: Enlarged Tonsils with pus on them

Symptoms of ARF are:

- Fever
- Painful and tender joints — most often the ankles, knees, elbows or wrists
- Pain in one joint that moves to another joint
- Red, hot or swollen joints
- Small, painless nodules beneath the skin
- Chest pain
- Sensation of rapid, fluttering or pounding heartbeats (palpitations)
- Fatigue or extreme tiredness
- Shortness of breath
- Flat or slightly raised, painless rash with a ragged edge that is round (erythema marginatum)
- Jerky, uncontrollable body movements — most often in the hands, feet and face (Sydenham Chorea)
- Outbursts of unusual behavior, such as crying or inappropriate laughing, that accompanies Sydenham chorea (uncontrollable movements)



Small, painless nodules under the skin



Sydenham chorea

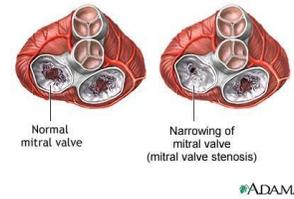


Rash in ARF

Pathophysiology of Rheumatic Heart Disease:

ARF does not often cause long term damage to the joints, or central nervous system. However, it can cause long term damage to the heart, causing Rheumatic Heart Disease. About 3 out of 5 people who get Acute Rheumatic Fever will develop Rheumatic Heart Disease (RHD). RHD is where some of the valves in the heart are damaged, most commonly the mitral valve and/or the aortic valve. Once someone has had rheumatic fever it is very easy for them to get it again, or to have another infection that could damage their heart more. It can be several years after an initial episode of Rheumatic Fever that valvular damage becomes evident.

Acute Rheumatic Heart Disease is characterized by inflammation in the layers of cardiac muscle. This inflammation causes a narrowing of the heart valves, usually the mitral valve or the aortic valve, called 'stenosis'. This means that the valve can't open fully and less blood can flow through the valve into the body.



This can cause 2 problems:

1. There will be too much blood inside the heart, causing a change in pressure, increased strain on the heart and the collection of fluid in the lung tissues
2. Not enough blood will reach the body – this can mean that oxygen is not delivered to cells. Body cells need oxygen in order to function properly

One of the most common problems in RHD is **Mitral Regurgitation**. A healthy heart is a closed system, where the blood only flows one way through valves and around the heart. MR is caused when the mitral valve does not close properly, allowing some of the blood to leak back through the valve and into the left atrium. **Aortic Regurgitation** is the same process, but involves the Aortic Valve instead of the Mitral Valve.

Symptoms of Rheumatic heart Disease:

Sometimes children who have RHD don't have any symptoms, and might not know that they are sick. The results from the ECG might be the only sign that there is something that is wrong with the child's heart. It is important to remember that the child does have a serious heart disease, and needs to be managed appropriately.

Symptoms of RHD could be:

1. Chest pain
2. Excessive fatigue
3. Heart palpitations
4. Shortness of breath

Diagnosis of Rheumatic Heart Disease:

All children who have had an episode of ARF in the last 10 years should be suspected of having Rheumatic Heart Disease. When it is suspected that a child has RHD, they should get the following tests.

1. *Echocardiogram*

RHD is diagnosed by an Echocardiogram (Echo). All children who have ARF, or who are suspected of having had an undiagnosed episode of ARF in the past should have an Echo. An echo will assess valvular function, and Left Ventricle functionality. An echo using ultrasound technology (like when women are pregnant and the doctor is checking on the baby) to assess valves, blood flow and pressure inside the heart. The following table shows what results you might see on a echo of a child who has RHD

Table 1: Expected Echocardiogram results in people with Rheumatic Heart Disease			
Aortic Stenosis			
	<i>Mild</i>	<i>Moderate</i>	<i>Severe</i>
Mean Gradient (mmHg)	Less than 25	25-40	More than 40
Valve Area (cm ²)	More than 1.5	1.0-1.5	Less than 1.0
Mitral Stenosis			
	<i>Mild</i>	<i>Moderate</i>	<i>Severe</i>
Mean Gradient (mmHg)	Less than 5	5-10	More than 10
Pulmonary Artery Systolic Pressure (PAPS)	Less than 30	30-50	More than 50
Valve Area (cm ²)	More than 1.5	1.0-1.5	Less than 1.0
Aortic Regurgitation			
	<i>Mild</i>	<i>Moderate</i>	<i>Severe</i>
Regurgitant volume (ml/beat)	Less than 30	30-59	More than 60
Regurgitant fraction (%)	Less than 30	30-49	More than 50
Left Ventricle Size	Normal	Normal	Increased
Mitral Regurgitation			
	<i>Mild</i>	<i>Moderate</i>	<i>Severe</i>
Regurgitant volume (ml/beat)	Less than 30	30-59	More than 60
Regurgitant Fraction (%)	Less than 30	30-49	More than 50
Left Atrial Size	Normal	Normal	Increased
Left Ventricle Size	Normal	Normal	Increased

2. Chest X-Ray

This will give an indication of the size of the heart, whether it is normal or enlarged.

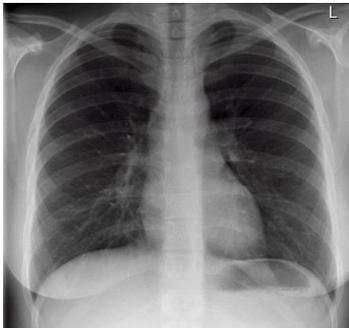


Figure 2: Normal Chest X Ray

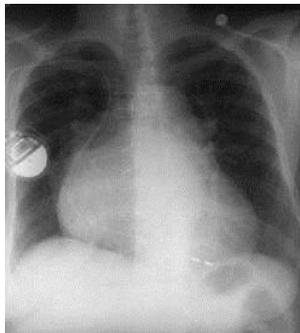


Figure 3: Enlarged Left Atrium



Figure 4: Enlarged Left Ventricle

3. Laboratory Tests

Antistreptolysin O (ASO, ASLO)

This test measures antibodies that are found when a child is infected with Rheumatic Fever. It can be useful to diagnose recent episodes of Rheumatic Fever, but does not diagnose RHD. ASO levels peak about 2 weeks after initial infection.

Complications of RHD:

Treatment and Management:

All cases of Strep Throat and Acute Rheumatic Fever need to be treated with antibiotics as soon as possible to prevent damage to the heart. Oral penicillin is an appropriate antibiotic to use in cases of Streptococcus infection. Patients with this infection should be very careful around others, because it is very contagious.

Once someone has been diagnosed with Rheumatic Heart Disease the focus changes from treatment to management and prevention of further damage. It is possible to reverse the damage caused by an episode of Rheumatic Fever, provided that there are no further instances of Streptococcal infection. Children who have a moderate to severe case of RHD, or those who get strep throat or ARF again don't have as good a prognosis, and often require further intervention, including surgery. There are several things that need to be done to manage children with Rheumatic Heart Disease.

1. Antibiotic Regime

This is the most important part of managing a child with Rheumatic Heart Disease. Current recommendations are management with a strict antibiotic regime to prevent further infection with the Streptococcus bacteria. All people who have a history of ARF or RHD should be receiving antibiotic therapy as secondary prevention. The most appropriate antibiotic is Penicillin. It is a broad spectrum antibiotic that is effective against Group A Streptococcus bacteria, is readily available in Vietnam and relatively affordable. Recent evidence shows that a monthly intramuscular injection is more effective at preventing re-infection with a Group A Streptococcal bacteria. However, this is rarely an appropriate route in Vietnam, as patients live far away from a medical facility. Oral therapy is the recommended antibiotic regime in Vietnam. Some people are allergic to penicillin, and so cannot take this medication (see table 2 for an alternative antibiotic). Symptoms of a Penicillin allergy in children are:

- Rashes or hives
- Itchy eyes
- **Swollen face, tongue or lips (this is very serious and if a child has this symptom, they need to see a doctor immediately).**

Table 2: Antibiotic Regime for Children with Rheumatic Heart Disease

Antibiotic	Dose	Route	Frequency
First line			
BPG (Penicillin)	BPG 900mg (1,200,000 units) >20kg BPG 450 mg (600,000 U) < 20 kg	Deep Intramuscular injection	4 weekly,

Second line (If im route is not possible, adherence should be carefully monitored)			
Phenoxymethylpenicillin (Penicillin V)	250 mg	Oral	Twice daily
Following documented penicillin allergy			
Erythromycin	250 mg	Oral	Twice daily

After the 10 years of antibiotic therapy has finished, the child will need another thorough assessment by a good cardiologist (a doctor who only works with hearts) or a doctor with experience with Rheumatic Heart Disease. If the child still has damage to their heart, they might need to stay on antibiotic therapy, or investigate other treatment options.

Table 3: Duration of antibiotic regimes

Status	Results	Action
All people with ARF or RHD	<ul style="list-style-type: none"> History of ARF Diagnosis of RHD 	Minimum 10 years after most recent episode of ARF* or until age 21 years (whichever is longer).
<i>After the 10 years of initial antibiotic therapy</i>		
No RHD	No pathological mitral or aortic regurgitation, but may have minor morphological changes to mitral or aortic valves on echocardiography	Discontinue at that time
Mild RHD	Mild mitral or aortic regurgitation clinically and on echocardiography, with no clinical evidence of heart failure, and no evidence of cardiac chamber enlargement on echocardiography	Discontinue at that time
Moderate RHD	<ul style="list-style-type: none"> Any valve lesion of moderate severity clinically (e.g. mild–moderate cardiomegaly and/or mild–moderate heart failure) or on Echo Mild mitral regurgitation, together with mild aortic regurgitation clinically or on echocardiography Mild or moderate mitral or aortic stenosis Any pulmonary or tricuspid lesion co-existing with a left heart valvular lesion 	Continue until 35 years of age
Severe RHD	<ul style="list-style-type: none"> Any severe valve lesion clinically (e.g. moderate to severe cardiomegaly or heart failure) or on echocardiography Any impending or previous valve surgery for RHD 	Continue until 40 years of age

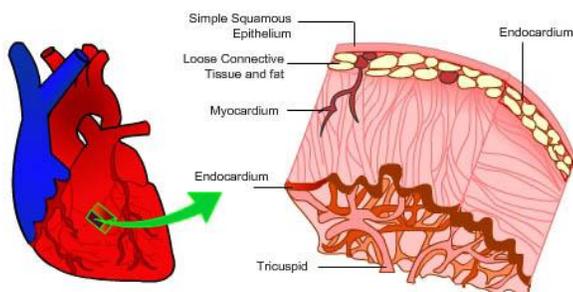
*This means that if a child catches ARF again after starting the antibiotic regime, they have to start from the beginning e.g. Nhi caught ARF when she was 11. She was put on an antibiotic regime of Penicillin V 250mg to have two times per day. The doctor told her she would need to take Penicillin V until she was 21 years old. When Nhi was 16, she caught ARF again. She went back to the doctor, who told her that now she needed to keep taking the Penicillin V twice per day until she was 26 years old.

2. Regular Echocardiograms and Follow up:

Echo's are an important part of managing a child who has RHD. Regular Echo's can show whether the child's heart disease is getting worse, or whether it is getting better. If the child's RHD is worsening, then treatment options need to be evaluated, and further medication therapy or surgical options should be discussed. A child with RHD should be getting Echo's every 6-12 months. Below is an easy guide to show how often children with RHD should be followed up.

Priority	Classification	Recommended Follow-up
Priority 1	<i>Severe RHD</i> Severe valvular disease or Moderate/severe valvular lesion with symptoms or Mechanical prosthetic valves, tissue prosthetic valves and valve repairs including balloon valvuloplasty	<ul style="list-style-type: none"> • An appropriate antibiotic regime • Clinic review every 3-6 months • Echo every 3-6 months • Dental review – within 3 months of diagnosis, then every 6 months • Flu vaccine every year • Endocarditis prevention as required
Priority 2	<i>Moderate RHD</i> Any moderate valve lesion in the absence of symptoms and with normal left ventricular function	<ul style="list-style-type: none"> • An appropriate antibiotic regime • Clinic review every 6 months • Echo every year • Dental review – within 3 months of diagnosis then every 6 months • Flu vaccine yearly • Endocarditis prevention as needed
Priority 3	<i>ARF with no RHD or Mild RHD</i> ARF with no evidence of RHD, or Trivial to mild valvular disease	<ul style="list-style-type: none"> • An appropriate antibiotic regime • Clinic review yearly • Echo every year • Dental review yearly

3. Prevention of Infective Endocarditis



Infective Endocarditis is a dangerous complication of Rheumatic Heart Disease. Infective Endocarditis is an inflammation of the inner tissue of the heart (also called the 'Endocardium') caused by a bacteria or a fungus. It is more common in people who have Rheumatic Heart Disease, and can cause significant damage to valves.

In Infective Endocarditis, the bacteria or fungus enters the bloodstream, usually from an invasive medical procedure. Most of the bacteria is killed by the immune system in the blood stream. But sometimes it can enter the heart, and cause a very serious infection. As the infection progresses, small clumps of bacteria or fungi or other debris from the infection form around heart valves. These clumps, called 'vegetations', prevent the valves from opening and closing properly. This affects the way that the heart pumps blood, and can cause heart failure.

Symptoms of Infective Endocarditis are:

- Fever – the fever is normally low grade (38 degrees to 39 degrees), and can come and go
- Petechiae – small red spots on the skin
- Osler nodes – tender nodules found under the skin, normally in the tips of the fingers
- Janeway lesions – non tender flat spots on the palms and soles of the feet
- Stiff neck
- Delirium
- Paralysis
- Unusual heart rhythms and unusual heart sounds (pericardial rub, pleural friction rub)
- Roth spots – haemorrhages in the eyes (this is very rare)

Infective Endocarditis is treated with antibiotics or antifungal medication. Early treatment will reduce the chance of significant and life threatening complications.

Prevention:

People who have Rheumatic Heart Disease are at increased risk of developing Infective Endocarditis. Therefore, it is very important to take preventative measures against this disease. The most recognized preventative method is providing people with RHD antibiotics before an invasive medical procedure. This way, if bacteria does enter the blood stream, it should be cleared from the body before it can make its way to the heart.

Procedures where people with RHD should receive prophylactic (preventative) antibiotics are:

Procedure	Appropriate antibiotic
Dental <ul style="list-style-type: none"> Dental extractions Dental surgery Replanting teeth If multiple procedures are being conducted, if the procedure will be a long time or periodontal disease is evident, antibiotic prophylaxis should be used for any dental procedure	Clindamycin (Child: 15 mg/kg up to 600 mg) 600 mg orally as single dose 1 hour prior to procedure or 600 mg iv, over at least 20 min just prior to procedure Vancomycin (Child less than 12 years: 30 mg/kg up to 1.5 g) 1.5 g iv by slow infusion, over at least 1 hour just prior to procedure Lincomycin (Child: 15 mg/kg up to 600 mg) 600 mg iv over 1 hour just prior to procedure Teicoplanin (Child: 10 mg/kg up to 400 mg) 400 mg iv just before the procedure or im 30 min before procedure
Respiratory Tract <ul style="list-style-type: none"> Tonsillectomy/adenoidectomy Bronchoscopy Surgery of the bronchial, sinus, nasal or middle ear 	Same antibiotic therapy as Dental procedures
Genitourinary and Gastrointestinal Tract <ul style="list-style-type: none"> Vaginal delivery with prolonged labour Any surgery where there is already an infection Incision/drainage of an abscess Any procedure through infected skin 	Vancomycin (Child less than 12 years: 30 mg/kg up to 1.5 g) 1.5 g iv by slow infusion, over at least 1 hour just prior to procedure Teicoplanin (Child: 10 mg/kg up to 400 mg) 400 mg iv just prior to procedure

4. Heart Medication

Children who develop severe valvular heart disease and become symptomatic need to be put on appropriate heart medication. There is a risk that children who have severe valvular lesions in RHD could develop clinical heart failure. In this case they will need to be placed on the following medication:

- Diuretics – to relieve fluid overload, swelling in the hands and feet, and fluid on the lungs
- Angiotensin-converting Enzyme (ACE) inhibitors – this drug opens blood vessels up, which allows more blood to flow through and decreases the work of an overloaded heart in moderate to severe Mitral or Aortic Regurgitation. Using this medication can reduce the need for surgery in some cases

5. Surgery

In cases of severe RHD, surgery might be necessary. It is more common for surgeons to recommend valve repair rather than valve replacement.

Table 5. Indications for surgery in advanced rheumatic valvular disease

Rheumatic Valvular Disease	Indications for Surgery	Choice of surgery
Mitral Regurgitation	<ul style="list-style-type: none"> • Patient is symptomatic – painful breathing, fatigue • Impaired Left Ventricular systolic function EF <60% • A very enlarged heart • Pulmonary hypertension (PAPS) >50mmHg • Atrial Fibrillation 	<ol style="list-style-type: none"> 1. Mitral Valve Repair 2. Mitral Valve Replacement – biological or mechanical*
Mitral Stenosis	<ul style="list-style-type: none"> • Patient is symptomatic – painful breathing, fatigue • Mitral Valve Area (MVA) <1.5 cm² • Pulmonary Hypertension (PAPS) >50mmHg 	<ol style="list-style-type: none"> 1. Percutaneous Balloon Mitral Valvuloplasty 2. Mitral valve repair or replacement (biological or mechanical)* if Mitral Valve is not suitable for option 1 (e.g. valve is heavily calcified) or if moderate or greater MR is present.
Aortic Regurgitation	<ul style="list-style-type: none"> • Moderate/severe AR with symptoms <p><i>Indications for surgery in patients with no symptoms:</i></p> <ul style="list-style-type: none"> • Left Ventricular function <55% • Significantly enlarged left ventricle 	<ol style="list-style-type: none"> 1. Bioprosthetic or homograft valve replacement 2. Mechanical Valve Replacement 3. Aortic Valve Repair 4. Ross procedure (replacement of the aortic valve with the pulmonary valve, and replacing the pulmonary valve with a homograft) this procedure should only be done in selected cases with experienced surgeons.
Aortic Stenosis	<ul style="list-style-type: none"> • Symptoms plus mean systolic gradient >40-50mmHg or Aortic Valve Area (AVA) <1.0 cm² • Impaired cardiac function (EF <50%) plus mean systolic gradient >40-50mmHg or AVA <1.0cm² 	<ol style="list-style-type: none"> 1. Bioprosthetic or homograft valve replacement 2. Mechanical valve replacement
Tricuspid Regurgitation	<ul style="list-style-type: none"> • Moderate/severe TR in association with symptomatic Mitral Valve Disease (MS or MR) • Progressive symptomatic right heart failure 	<ol style="list-style-type: none"> 1. Tricuspid valvuloplasty 2. Tricuspid valve replacement with mechanical or biological valve if valvuloplasty not available
Tricuspid Stenosis	<ul style="list-style-type: none"> • Moderate/severe TS in association with symptomatic Mitral Valve Disease (MS or MR) • Progressive right heart failure 	<ol style="list-style-type: none"> 1. Percutaneous balloon valvuloplasty or surgical commissurotomy 2. Tricuspid valve replacement with mechanical or biological valve if option 1 not available

A note on valve replacement surgery:

There are many factors that need to be considered when deciding which surgery a patient should have. When choosing between a mechanical valve or a bioprosthetic/homograft valve, it is important to consider the following:

Mechanical Valve	Bioprosthetic/homograft valve
<ul style="list-style-type: none"> • Lasts for a long time 	<ul style="list-style-type: none"> • Does not last as long so might need

	another surgery
<ul style="list-style-type: none">• Patient needs to be on anticoagulation therapy, everyday for the rest of their life<ul style="list-style-type: none">• If the patient is female, and planning on having children, it is better for them to have a bioprosthetic/homograft valve• If the patient is not good at taking their medicine, it can put them at risk of developing a blood clot	<ul style="list-style-type: none">• Does not need anticoagulation therapy