The cover of the guidelines features a vibrant red background with a complex, repeating geometric pattern in a lighter red shade. The pattern consists of various shapes including diamonds, zig-zags, and stylized floral motifs. The title is centered in white text.

# Fiji Guidelines for Acute Rheumatic Fever Fever and Rheumatic Heart Disease Diagnosis Management and Prevention

## Evidence Based Best Practice Guidelines



## Scope and Purpose

These guidelines have been developed by an expert writing group (which included infectious disease, cardiology, epidemiology and public health practitioners) to aid the diagnosis and clinical management of Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease (RHD) in Fiji. The guidelines have been based on international best practice guidelines for ARF/ RHD including: the Jones criteria update 2015[1], The New Zealand Guidelines for Rheumatic Fever 2014[2], The Australian Guidelines for Prevention, Diagnosis and Management of Rheumatic Fever and Rheumatic Heart Disease 2012[3], the Fiji Cardiovascular Guidelines 2015, the Fiji Obstetrics and Gynaecology Clinical Practice Guidelines 2015, the Fiji National RHD Policy 2015, the Fiji Ministry of Health and Medical Services National Strategic Plan 2016-20, the World Heart Federation Diagnosis and Management of Acute Rheumatic Fever Rheumatic Heart Disease 2008[4] and the WHO Expert Consultation Technical Report: Rheumatic Fever and Rheumatic Heart Disease 2003.[5]

The purpose of these guidelines is to:

- identify the evidence for best practice in ARF and RHD diagnosis,
- identify the standard of care that should be available for people in Fiji; and
- provide an evidence based protocol for diagnosis, treatment and prevention of ARF and RHD in Fiji.

## Disclaimer

The authors do not warrant the accuracy of the information contained in these guidelines and do not take responsibility for any deaths, loss, damage or injury caused by using the information contained herein. While every effort has been made to ensure that the information contained in these guidelines is correct and in accordance with current evidence based clinical practice, the dynamic nature of medicine requires that users in all cases employ independent professional judgment when using these guidelines.

## Funding support

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## **Foreword**

The Fiji RHD Prevention and Control Program is established within the Wellness Centre of the Ministry and Medical Services. The program works to improve the diagnosis, management and reporting of Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease (RHD) for the people of Fiji.

This set of ARF and RHD guidelines have been developed using evidence based resources locally and internationally, in collaboration with key stakeholders and experts. The guidelines are intended to be used by nurses, doctors and allied health professionals to assist in the provision of standardised care and optimal patient management. The guidelines are accompanied by a summary guideline and ARF clinical diagnostic posters.

The Ministry will continue to engage with other ministries and donor partners, in promoting healthy lifestyles and behaviour changes in communities to reduce the burden of ARF and RHD and at the same time consolidate knowledge and practice of health professionals. It is our hope that these guidelines will be used effectively by Fijians to gain knowledge and skills to equip them to perform their roles very well in the communities they serve.

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## List of Abbreviations

AF	Atrial Fibrillation
AHA	American Heart Association
AMVL	Anterior Mitral Valve Leaflet
ARF	Acute Rheumatic Fever
ASOT	Anti-streptolysin Otitre
AV	Aortic Valve
BPG	Benzathine Penicillin G
CHD	Congenital Heart Disease
CWMH	Colonial War Memorial Hospital
CXR	Chest X ray
ECG	Electrocardiogram
EPI	Expanded Programme for Immunisation
ESD	End Systolic Dimension
ESR	Erythrocyte Sedimentation Rate
FBC	Full Blood Count
GAS	Group A Streptococcus
GBD	Global Burden of Disease
GrASP	Group A Streptococcal Project (research group)
HIU/HIS	Health Information Unit/ Health Information System
IM	Intramuscular
IU	International Units
JVP	Jugular Venous Pressure
LFT	Liver Function Tests
LVEDD	Left Ventricular End Diastolic Dimension
LVEF	Left Ventricular Ejection Fraction
LVESD	Left Ventricular End Systolic Dimension
LVESV	Left Ventricular End Systolic Volume
LMWH	Low Molecular Weight Heparin
MoHMS	Ministry of Health and Medical Services
MFAT	Ministry of Foreign Affairs and Trade, New Zealand
MV	Mitral Valve
NYHA	New York Heart Association
PHIS	Public Health Information System
PMVL	Posterior Mitral Valve Leaflet
RFIS	Rheumatic Fever Information System
RHD	Rheumatic Heart Disease
U and E	Urea and Electrolytes
UFH	Unfractionated Heparin
WHF	World Heart Federation
WHO	World Health Organization
YLL	Years of Life Lost

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## **Section 1: Guidelines and clinical pathways for ARF**

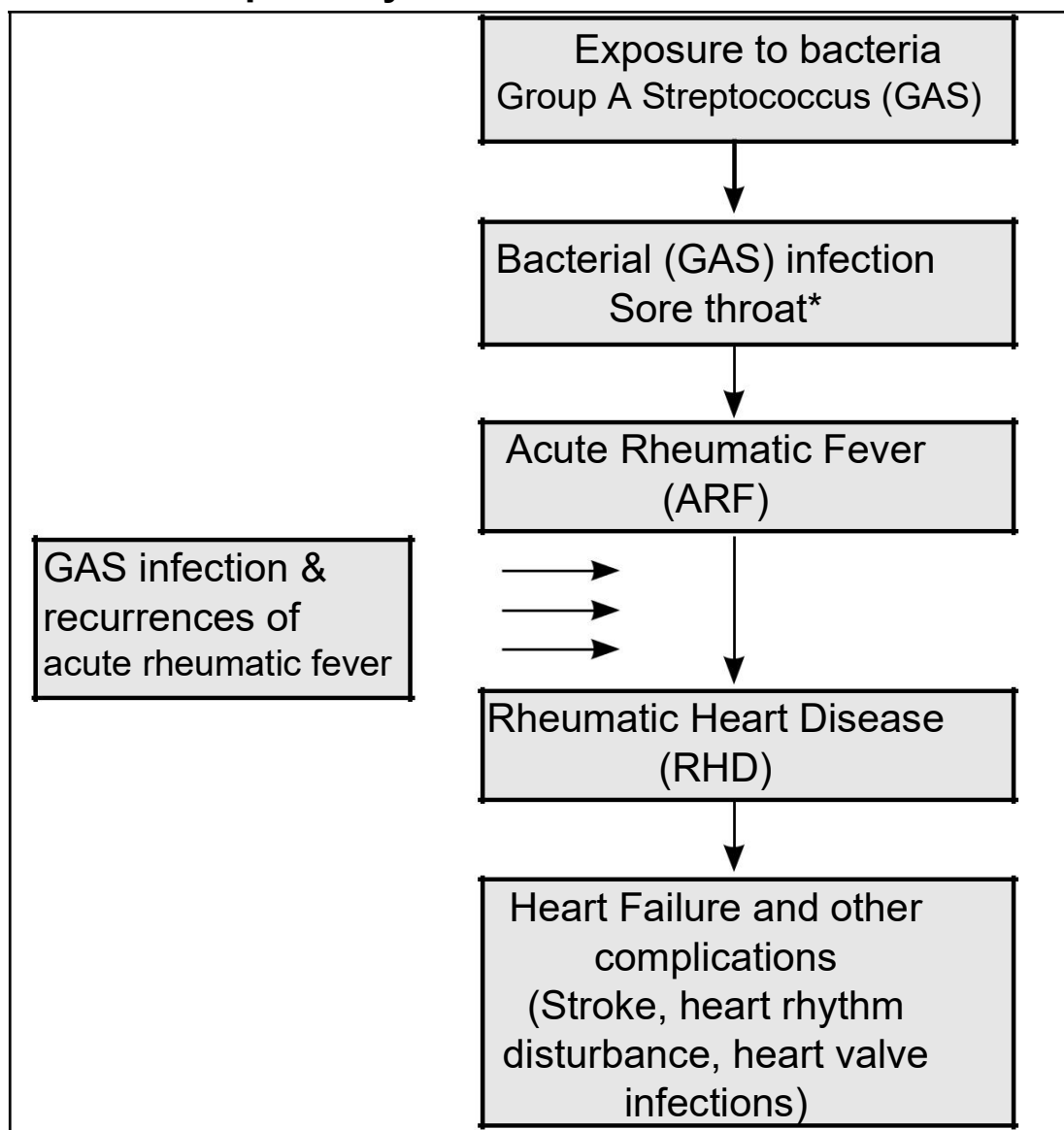
### **Acute Rheumatic Fever Key Messages**

- ARF occurs following a group A streptococcal (GAS) infection
- The incidence of ARF in Pacific Islanders is amongst the highest in the world
- ARF predominantly affects children aged 5 to 15 years
- ARF and RHD largely affect disadvantaged populations
- Crowding in the household and poverty are associated with an increased risk of developing rheumatic fever
- There is no convincing evidence of a genetic cause of rheumatic fever and no reliable genetic markers of susceptibility to the disease. Historically ARF has affected all races in all parts of the world.
- Accurate diagnosis of ARF requires a combination of clinical criteria, laboratory and echocardiographic investigation
- Diagnosis ideally requires hospital admission for accurate diagnosis and optimal management
- A first attack of acute rheumatic fever is potentially preventable if the individual presents with GAS pharyngitis and receives effective antibiotic treatment
- There is some limited evidence that rheumatic fever is caused by skin infections, so treating skin infections is also considered important in the prevention of acute rheumatic fever
- ARF patients are at 10 times the risk for subsequent episodes of ARF
- Delivery of regular Benzathine penicillin G injections to ARF cases is the mainstay of secondary prevention, preventing recurrence of ARF and progression of RHD.

## What is Acute Rheumatic Fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection by group A streptococcus (GAS). It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF generally leaves no lasting damage to the brain, joints or skin, but can cause persisting and life threatening heart damage, which is then called rheumatic heart disease (RHD). (Box 1)

### Box 1 Causal pathways for ARF and RHD



## **1.1 Epidemiology**

### **Global Epidemiology**

It is well established that during the 20th century, the incidence of ARF and the prevalence of RHD declined substantially in Europe, North America, and other developed nations. This decline has been attributed to improved hygiene, improved access to antibiotic drugs and medical care, reduced household crowding, and other social and economic changes.[5] Although sporadic cases of ARF continue to be seen in affluent nations, the major burden is currently found in low- and middle-income countries and in selected Indigenous populations.[6]

### **Who gets ARF?**

Acute rheumatic fever is seen predominantly in children aged 5-14 years, although recurrent episodes may occur in people aged into their 40s. Rheumatic Heart Disease represents the cumulative heart damage caused by previous ARF episodes. The prevalence of RHD peaks in the third and fourth decades of life.

### **Fiji Epidemiology**

Data on ARF and RHD in Fiji were not routinely recorded until the establishment of a national RHD Register and commencement of the World Heart Federation RHD control programme in 2005. Since this time, activity around RHD has increased and a number of epidemiological and clinical research studies undertaken by the Fiji Group A Streptococcal Project (Fiji GrASP) have provided an insight into the burden of ARF and RHD in Fiji.

### **ARF incidence**

In Fiji estimates of the incidence of first episodes of ARF in children and young adults range from 15 to 25 per 100,000 per year.[7, 8] Many children present late with valve damage (RHD) because many ARF presentations are not recognized in Fiji.[9] The researchers found, “patients presenting with potential features of ARF seldom had a diagnostic evaluation sufficient to exclude its diagnosis” suggesting that many clinical staff working in the high incidence setting of Fiji are not familiar with the symptoms of ARF.[7]

For example, if no murmur is heard on auscultation or if auscultation is not performed, diagnosis of ARF or RHD may be dismissed.

Similarly, if there is no presenting joint involvement with ARF, a diagnosis can be dismissed.[10] In Fiji patients frequently present late with advanced RHD and significant valvular damage.

## **RHD prevalence**

Nearly 1% of all Fijians have evidence of RHD (10,11) with echocardiographic confirmed prevalence of RHD in school-aged children in Fiji estimated at 8.4 per 1000[11] and the Global Burden of Disease 2010 study estimated an all age prevalence of RHD of 9.8 per 1000.[12] The Indigenous I-Taukei Fijians are more likely to have RHD than the Indo-Fijian and RHD is more prevalent in women.[12]

## **RHD mortality**

Recent studies focusing on RHD mortality have found that death from RHD is common (2.4% of autopsy cases audited)[13] and that significant numbers of young people die from RHD in Fiji with a mean age of death of 38 years.[12] Age-standardised death rates from RHD are more than twice those reported in current global estimates. In a study using record linkage in Fiji it was found that between 2008 and 2012 there were 378 deaths attributable to RHD, with over half occurring in people aged less than 40 years.[14] In this study RHD was the second most common cause of death in people aged 5 – 29 years, behind drowning. RHD leads to 9.9 deaths (95% CI 9.8–10.0) and 331 years of life-lost (YLL, 95% CI 330.4–331.5) per 100,000 person-years.[15] Thus, RHD is a leading cause of premature death in Fiji.[14]

## **Cost to Fiji**

RHD is costly to the Fiji health system. The estimated annual total cost of RHD in Fiji is US\$6,077, 431. The largest cost item comprised productivity losses from premature mortality (77% of total cost of illness). The total indirect costs of RHD were 20 times larger than the total direct costs of health care.[14]



## 1.2 Rheumatic Fever Prevention

Interventions on the causal pathway from Group A Streptococcal infection to chronic RHD are typically considered in four areas of intervention. (Box 2)

### Box 2: Intervention opportunities for ARF/ RHD

<b>Primordial Prevention</b>
Reducing household overcrowding, poverty and malnutrition Improved access to health care
<b>Primary Prevention</b>
Treating sore throats due to GAS with antibiotics Development of a vaccine against GAS
<b>Secondary Prevention</b>
ARF/RHD register The continuous administration of penicillin for those who have had ARF and for those with RHD
<b>Tertiary Intervention</b>
Medical management of severe RHD Heart valve surgery

### Primordial prevention

Primordial prevention is key to reducing the incidence of ARF and RHD. Household overcrowding, poor sanitation and reduced nutrition and living conditions have all been shown to contribute to increasing the risk of ARF/RHD.[16-19]Over the past century ARF has largely disappeared from middle to high-income countries and now exists primarily in resource-poor settings. Improving social determinants of health can lead to reduction in ARF/RHD incidence. Targeted measures undertaken at an individual household or community level may also have an impact in reducing the risk of recurrence of ARF.

## Primary Prevention

Primary prevention is a strategy that seeks to prevent disease occurring in the first instance rather than treating it once it has developed. In the case of ARF this means treating Group A streptococcus bacterial throat infections before they can initiate ARF. When an individual is exposed to GAS, the organism attaches to and colonises the pharyngeal mucosa. A process of infection incorporating an immune response is initiated, and an episode of ARF may occur 2–3 weeks later. The aim of primary prevention is to identify symptomatic GAS pharyngitis (sore throats) in those individuals most at risk of ARF (typically children aged 5–14 years), and treat and eradicate the bacteria with antibiotics. Studies show that ARF associated with GAS pharyngitis can be prevented if treatment is commenced within 9 days of symptoms appearing. While the association between GAS pharyngitis and ARF is well described, the role of GAS-associated skin infection remains unclear.[20] Most experts would advise treatment of GAS skin infections with antibiotic therapy, to achieve cure of the infection and to potentially reduce the risk of subsequent ARF.

## Antibiotic treatment of sore throats

Prevention of an initial attack of ARF requires the prompt and accurate diagnosis and adequate antibiotic treatment of GAS throat infections. ARF can be prevented if the preceding throat infection is treated in a timely and effective way. Recommended treatment of streptococcal throat infection is intramuscular (IM) benzathine penicillin or a ten-day course of oral (twice a day) phenoxymethyl penicillin or once a day amoxicillin which eradicates the streptococci from the pharynx. The role of the treatment of impetigo (skin sores) in the control of rheumatic fever is less well established, however Fiji has a very high incidence of skin disease and treatment for skin disease has been included in the accompanying *Fiji Ministry of Health and Medical Services (MoHMS) Sore Throat and Skin Sore Guidelines*.

People who have had an episode of acute rheumatic fever are at high risk of a subsequent episode. The Fiji RHD programme technical Advisory Group suggests a housing assessment and targeted education for individuals who have had an episode of ARF or have RHD. This should include an assessment of hygiene, nutrition and sanitation. For example the availability of soap, water, washing facilities for bodies and clothes – ‘clean house clean body message’ and good nutrition. Patient and family education is important, with a focus on reducing risk of recurrence and increasing adherence to secondary prophylaxis:

- Reduce numbers of people sleeping in same room
- Treat suspected bacterial sore throats, scabies and skin sores
- Present early to clinic to prevent complications
- Importance of regular BPG for those individuals with a history of ARF or RHD

**Detailed guidelines for Primary Prevention and the management of acute sore throat and skin disease in Fiji are provided in a separate guideline.**

This guideline provides clinical guidance for clinicians faced with the patient presenting with acute sore throat as their primary complaint including around the role of clinical decision rules, the role of throat swabbing, choice of antibiotic treatment and appropriate follow-up. [21-23]

## **Secondary Prevention**

**Secondary prevention of further episodes of ARF is of the highest priority for best practice care.**

Secondary prophylaxis with regular benzathine penicillin G (BPG) is the only RHD control strategy shown to be effective and cost-effective at both community and population levels. The appropriate duration of secondary prophylaxis is determined by age, time since the last episode of ARF and potential harm from recurrent ARF, but is likely to be 10 years or more. While secondary prophylaxis is a proven strategy for controlling RHD, and is also simple, cheap and cost-effective, it must be adequately implemented. Persistent high rates of recurrent ARF in high-risk populations highlight the continued barriers to secondary prevention.

These factors relate to overcrowded housing, poor access to health services, limited educational opportunities and poor environmental conditions.

Communities with the highest rates of ARF and RHD are often the least equipped to deal with the problem. Secondary prevention should include:

- BPG injections to provide continuous delivery of penicillin
- Strategies aimed at optimizing the delivery of secondary prophylaxis and patient care
- Nursing and medical staff to deliver the BPG on time
- A well-coordinated register based system for monitoring and reporting secured availability of penicillin
- The provision of education for, and coordination of available health services
- Community advocacy and health promotion activities
- Appropriate clinical and diagnostic resources and guidelines

## **Tertiary prevention**

Tertiary prevention activities aim to avoid early mortality and include the medical management of symptomatic RHD, anti-coagulation, triage of candidates for cardiac surgery and delivery of cardiac surgery. Tertiary prevention does not have an impact of the incidence of disease and will not assist to control ARF/ RHD at a population level. However, optimal care for those with RHD reduces the burden of living with RHD, by controlling symptoms and extending life. Improving clinical care will maximize the benefit of surgery (if access to cardiac surgery is available) by ensuring the most suitable candidates are selected for intervention and capacity is strengthened for the delivery of secondary prevention and anti-coagulation medications. Surgical intervention is tremendously expensive, does not 'cure' the underlying disease, nor prevent the development of new cases in high risk communities. While there is a clear role for cardiac surgery in RHD to alleviate symptoms and prevent heart failure, the best approach to minimise surgical risk and maximise social benefit is yet to be clearly defined in the Pacific region.

## 1.3 Acute Rheumatic Fever Diagnosis

### Summary of key diagnostic requirements

- The diagnosis of ARF requires health professionals to have a high index of suspicion and to be aware of the diagnostic criteria. Hospital referral where expertise is available for accurate diagnosis particularly echocardiography, is usual.
- It is important that an accurate diagnosis of ARF is made as:
  - A misdiagnosis may result in the individual receiving benzathine penicillin G (BPG) injections unnecessarily every four weeks for a minimum of 10 years.
  - Under-diagnosis of ARF may lead to the individual suffering a further attack of ARF, further cardiac damage and premature death.
- There is no single laboratory test specifically diagnostic for ARF; diagnosis is based on full clinical assessment and assessment of the probability of ARF. Clinicians experienced with ARF can play a key role to help make this decision. The probability for a diagnosis of ARF varies according to location and ethnicity. e.g. in a region with high incidence of ARF, such as Fiji, a person with fever and arthritis is more likely to have ARF than one in a low incidence region.
- The American Heart Association Jones diagnostic criteria for ARF have evolved over many years. The 2015 Revision incorporates Doppler echocardiography and brings the AHA into closer alignment with other international guidelines (that have been using Doppler echocardiography to its fullest extent for over 20 years). [1]
- In all cases, every attempt should be made to follow the criteria, including demonstrating evidence of GAS infection and using echocardiogram, but where this is not possible, a clinical diagnosis of probable ARF may be made by an experienced clinician.
- All patients with suspected or definite ARF (first episode or recurrence) should undergo echocardiography within 6 months (if not immediately available) to identify evidence of carditis and categorised as Probable ARF, until echocardiography confirmation is available to accurately classify the case.

Where possible all patients with suspected ARF (first episode or recurrence) should be hospitalised as soon as possible after the onset of symptoms. This ensures that all investigations are performed, and if necessary, the patient should be observed to confirm the diagnosis before commencing treatment. Hospitalisation also provides opportunity for patient education and counselling especially regarding the need for secondary prophylaxis.

## **Problems with diagnosis and management**

A number of factors contribute to the barriers in diagnosis and management of ARF in Fiji. These include:

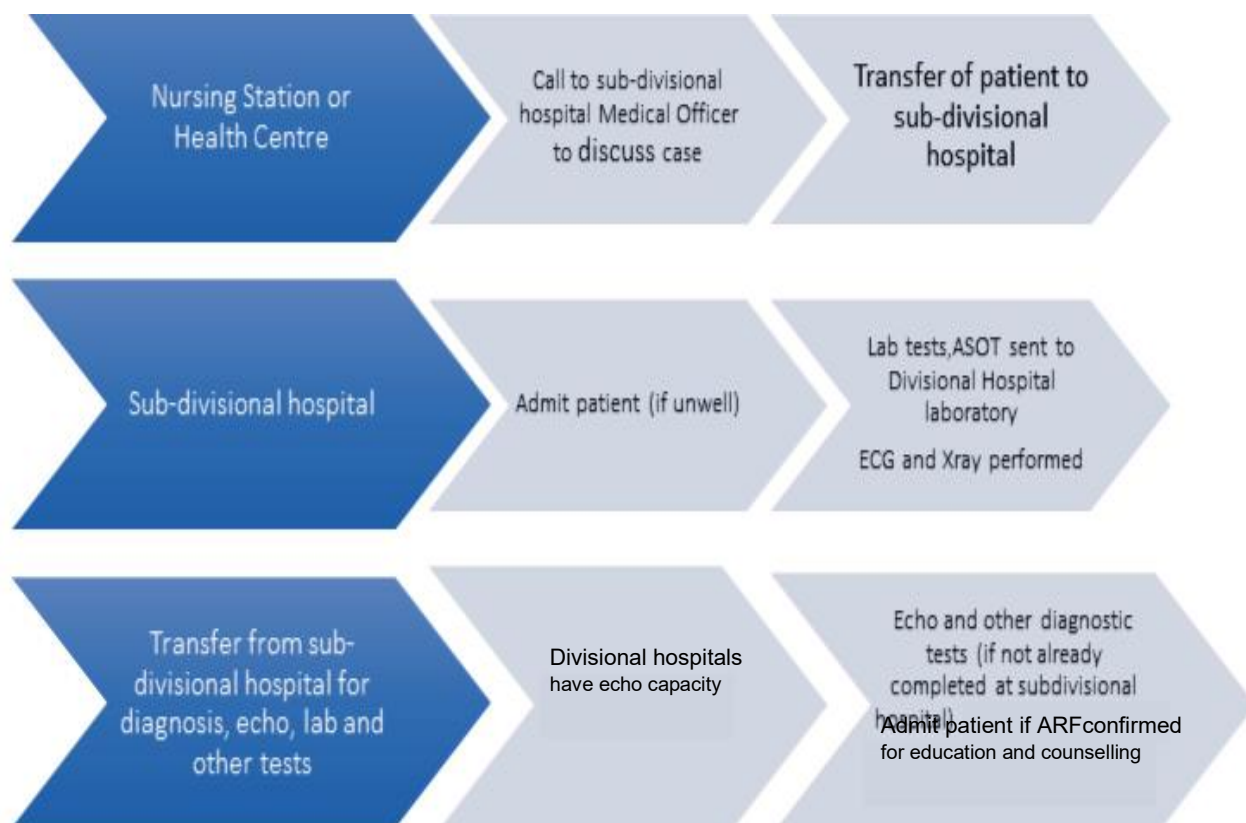
- The timely and accurate diagnosis of first ARF episode
- Health seeking behaviour
- Delivery of secondary prophylaxis medication to reduce risk for subsequent ARF episodes
- Many clinicians may have not seen cases of ARF
- Patients face transportation cost issues in accessing specialist diagnosis and care; and
- Access to laboratory tests and echocardiography is limited outside the major population centres.

## **Referral pathways for ARF**

Labasa Hospital and a number of sub-divisional hospitals are serviced by visiting clinicians from Suva. Therefore referral pathways for diagnosis for ARF vary across the country. The two main referral pathways in Fiji are:

1. Health clinic referral to sub-divisional hospital for laboratory and Senior Medical Officer confirmation (echocardiography to be booked with visiting echo team on rotation)
2. Health clinic/ sub-divisional hospital to Divisional Hospital Urban and peri-urban i.e. near CWM, Labasa and Lautoka Hospitals – Medical Officer refers to cardiologist/paediatrician and/or echo clinic. See Figure 1.

**Figure 1: Referral pathways for suspected ARF cases**



### Protocol for referral

- At the health centre or nursing station level the medical officer or nurse practitioner should refer the patient with suspected ARF to the nearest sub-divisional hospital for investigation. Investigations should be for all:
  - Laboratory tests: FBC, LFT, U and E, ESR, ASOT
  - Cardiac tests: ECG and CXR
  - Throat swab (if possible).
- Ideally the patient remains in hospital until the results are back i.e. ARF cannot be diagnosed until ASOT is back.
- A specific “ARF referral form” should be completed and the senior medical officer at the sub-divisional hospital contacted by phone to discuss the referral.
- All patients with suspected ARF should be discussed with someone expert in ARF (a paediatrician/physician/ cardiologist) and a plan for referral and admission should be arranged.

- All unwell patients presenting in heart failure\* or with carditis, must be admitted and evacuated to a Divisional Hospital. This may involve consultation with SMO/Cardiologist/Paediatrician/ Physician.
- Patients presenting at Divisional hospitals will be reviewed by a specialist/ Cardiologist as early as possible and investigated accordingly.

**\*Heart failure is defined by breathlessness walking or at rest, resting tachycardia, as well as classical signs of raised jugular venous pressure (JVP), hepatomegaly and crackles.**

The minimal list of tests for diagnosis that should be available at a Sub-divisional hospital is included in Table 1 below.

**Table 1: Minimal list of tests at Sub-divisional hospital level**

Tests to be undertaken at Sub-divisional hospital	Refer to divisional hospital or visiting specialist team
Clinical history	
Clinical examination	
Lab tests: WBC, ESR, ASOT*	Referral to divisional hospital for echocardiography or arrange with MoHMS specialist visiting team to
ECG	echo within 3 months
CXR	
Throat swab (if possible)	

*\*ASOT may not be currently available at all sub-divisional hospitals. If ASOT not available blood sample should be sent to the nearest Divisional hospital*

## **Diagnostic criteria for ARF**

Diagnosis of ARF is based on a combination of clinical criteria laboratory investigation and echocardiography. As of 2015, some sub-divisional hospitals in Fiji are equipped to undertake laboratory investigations, such as ASOT, however the majority do not have this facility. The diagnosis for ARF in Fiji is guided by the 2015 Revision of the Jones Criteria.[1] The Fiji Criteria for the diagnosis of ARF are presented in Table 2 and investigative tests are presented in Box 3. Echocardiogram is available by referral at all divisional hospitals.



## Table 2: Fiji Criteria for ARF Diagnosis[1]

	Criteria
Diagnosis: initial ARF	2 major manifestations <u>OR</u> 1 major plus 2 minor manifestations <u>PLUS</u> evidence of GAS infection*
Diagnosis: recurrent ARF	2 major <u>OR</u> 1 major plus 2 minor <u>OR</u> 3 minor <u>PLUS</u> evidence of GAS infection*
Major manifestations	<ol style="list-style-type: none"> <li>1. Carditis <i>-Clinical and/or sub-clinical (on echocardiography)</i></li> <li>2. Arthritis <i>-Monoarthritis, polyarthritis or polyarthralgia</i></li> <li>3. Chorea**</li> <li>4. Erythema marginatum</li> <li>5. Subcutaneous nodules</li> </ol>
Minor manifestations	<ol style="list-style-type: none"> <li>1. Monoarthralgia (unless arthritis is included as a major manifestation)</li> <li>2. Fever (<math>\geq 38^{\circ}\text{C}</math>)</li> <li>3. ESR <math>\geq 30</math> mm/h or white cell count <math>\geq 15 \times 10^6/\text{mL}</math></li> <li>4. Prolonged P-R interval, for age (unless carditis is a major manifestation)</li> </ol>

\* Evidence of preceding group A streptococcal infection is defined by at least one of the following:

1. Elevated ASOT above cut-offs by age determined for Fiji (see Table 3)
2. A rising ASOT defined as a twofold or greater difference between titres measured at presentation and when convalescent (2-4 weeks later generally)
3. A positive throat swab for group A streptococcus at presentation

\*\* Note that ARF can be diagnosed on the basis of chorea without other manifestations or evidence of GAS infection.

## ARF categories

### 1. ARF

Fulfil Fiji diagnostic criteria (Table 2)

Note that ARF can be diagnosed on the basis of chorea without other manifestations or evidence of GAS infection.

## **2. Probable ARF:**

In some circumstances, a clinical presentation may not fulfil the full ARF criteria (Table 2), yet the clinician may still have good reason to suspect that ARF is the diagnosis. In such situations, clinicians should use their discretion and clinical acumen to make the diagnosis that they consider most likely and manage the patient accordingly. A senior clinician should be involved in this decision.

This may occur where, for example:

- Testing for evidence of GAS infection (ASOT or throat swab) is unavailable or results pending
- Testing for ESR and WCC is unavailable
- Echocardiography is unavailable
- The history is not considered reliable or documentation of clinical features is not clear.

Specific clinical scenarios where a diagnosis of “Probable ARF” could be considered

- Carditis on echocardiography with only 1 minor manifestation
- Arthritis or polyarthralgia with 0 or 1 minor manifestations. Note that migratory polyarthritis or migratory polyarthralgia affecting the large joints is highly suggestive of ARF.

## Box 3 : Investigations in Suspected ARF

### Recommended for all cases

- White blood cell count
- Erythrocyte sedimentation rate (repeat weekly once diagnosis confirmed)
- Blood cultures if febrile (and where available)
- Electrocardiogram (ECG) repeat as necessary if conduction abnormality more than first degree)
- Chest x-ray
- Echocardiogram (repeat as necessary in 2-4 weeks if equivocal, or if severe carditis)
- **Throat swab (preferably before giving antibiotics)** culture for group A streptococcus
- Anti-streptococcal serology: anti-streptolysin O, if available (repeat 10-14 days later if first test not confirmatory)

### Tests for alternative diagnosis depending on clinical features:

- Repeated blood cultures if possible to exclude endocarditis or septic arthritis
- Joint aspirate (microscopy and culture) for possible septic arthritis\*
- Joint X-ray
- Dengue Fever rapid antigen and Elisa tests, arboviral testing, Chickungunya, Zika Virus (if available)
- Serology and autoimmune markers for autoimmune or reactive arthritis (including ANA - Anti Nuclear Antibody) - initial screening can be done at CWMH

\* Typically, the synovial fluid in joints affected by ARF contains 10,000 to 100,000 white blood cells/mm<sup>3</sup>. In septic arthritis the white cell count may be higher, bacteria may be seen on Gram stain and the culture for bacteria (usually *Staphylococcus aureus*) is positive. ASOT testing is not widely available in Fiji outside the Divisional hospitals. Patients presenting to sub-divisional hospitals will require a blood sample for ASOT to be taken and sent to the nearest Divisional hospital.

## Evidence of preceding Group A Streptococcal infection

In Fiji, evidence of preceding group A streptococcal infection is defined by at least one of the following:

- Elevated ASOT above cut-offs by age determined for Fiji (see Table 3)
- A rising ASOT defined as a fourfold or greater difference between titres measured at presentation and when convalescent (2-4 weeks later generally)
- A positive throat swab for Group A streptococcus at presentation

If the initial ASOT level is above the age specific cut-off this is sufficient to confirm diagnosis. If the initial ASOT level is below the age specific cut-off, ASOT should be repeated in 2 weeks.

**Table 3: Upper limits for serum antibody titres**

Age group (years)	UNL (U/ml) ASO Titre
1-4	170
5-14	276
15-24	238
25-34	177
≥ 35	127

## **1.4 Clinical Features of Acute Rheumatic Fever**

### **1.41 Major manifestations**

#### **Joint pain**

- Joint pain is the most common presenting symptom of ARF (occurring in up to 75% of first attacks).

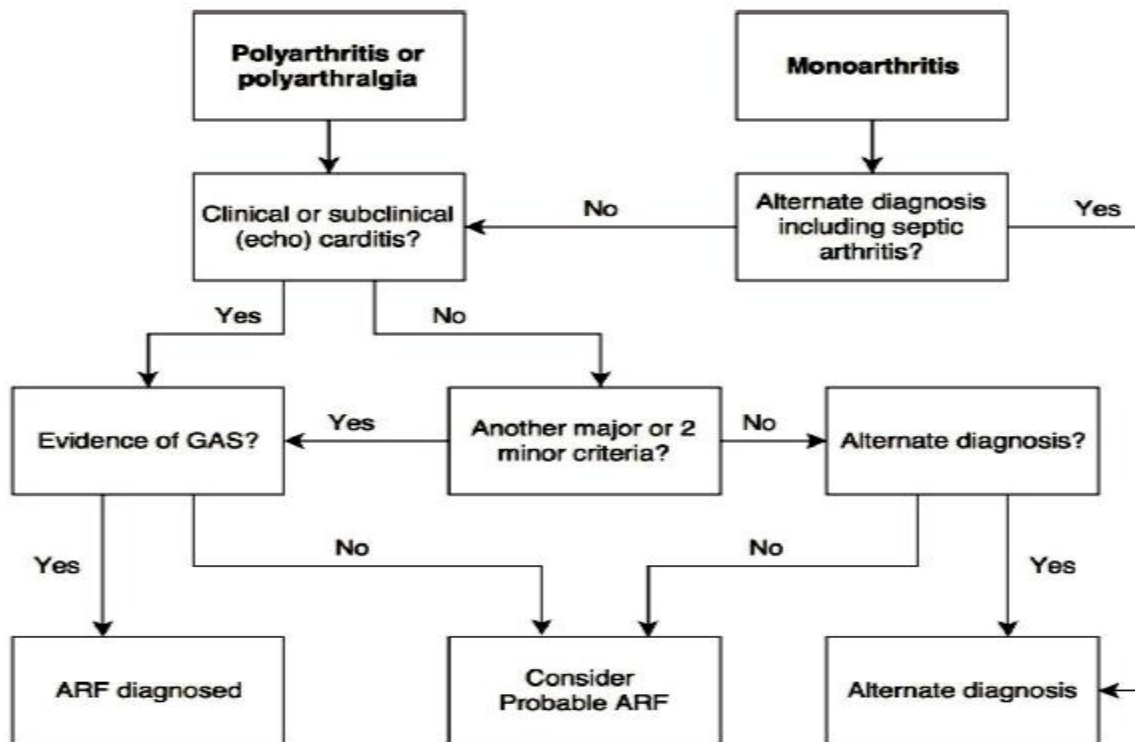
#### **Classic polyarthritis of ARF**

- Arthritis is defined as swelling of the joint in the presence of two or more of the following: limitation of movement, hotness of the joint, pain in the joint or localized tenderness.
- The classic arthritis of ARF is asymmetrical large joint (especially knees and ankles), polyarthritis (multiple joints) that is migratory (one joint becoming inflamed as another subsides) or additive (multiple joints progressively becoming inflamed without waning).
- Classic ARF arthritis as described above can be considered as ARF in high incidence settings unless proven otherwise.
- The arthritis of ARF is extremely painful, often out of proportion to the clinical signs. The arthritis of ARF is especially susceptible to aspirin and other non-steroidal anti-inflammatory drugs, usually responding within 24 hours of commencement of treatment and almost always within 48-72 hours.
- Because of the migratory nature of the arthritis, a definite history of arthritis, rather than documentation by the clinician, is sufficient to satisfy this criterion.
- History of a painful hip too painful to walk is accepted as evidence of arthritis of a hip.

### ***Mono-arthritis and polyarthralgia***

- In high RHD prevalence regions such as the Pacific Islands, the classic polyarthritis of ARF is not always observed, and mono-arthritis (arthritis of a single joint) or polyarthralgia (pain in multiple joints without evidence of inflammation) may be the presenting feature of ARF.
- As such, either mono-arthritis or polyarthralgia can be considered as a major manifestation of ARF in Fiji.
- In patients with possible ARF that present with arthritis of a single joint only, some experts recommend initial treatment with paracetamol or codeine for pain relief until a second joint is involved so that the classic pattern of ARF arthritis is observed, thereby facilitating the diagnosis of ARF.
- It is important to rule out septic arthritis in the patient who looks toxic and has a mono-arthritis. In the hospital setting, physicians and surgeons should collaborate when the diagnosis of arthritis is unclear. The diagnosis of a single swollen joint with a high fever is septic arthritis unless joint aspirate negative. Patients with sterile joint aspirates should never be treated speculatively for septic arthritis without further investigation, particularly in areas with a high ARF/RHD prevalence, such as Fiji. (18-20).
- Arthralgia differs from arthritis in that there is pain on joint movement without evidence of swelling or heat. It is a non-specific symptom.
- Arthralgia involving multiple joints may suggest ARF if it appears in the same pattern as rheumatic polyarthritis: that is, asymmetrical, affecting large joints, and migratory or additive.
- Mono-arthritis and polyarthralgia are often associated with overt or subclinical carditis. [24]
- ARF should always be considered in the differential diagnosis of patients presenting with joint pain in high-risk populations (Algorithm 1).

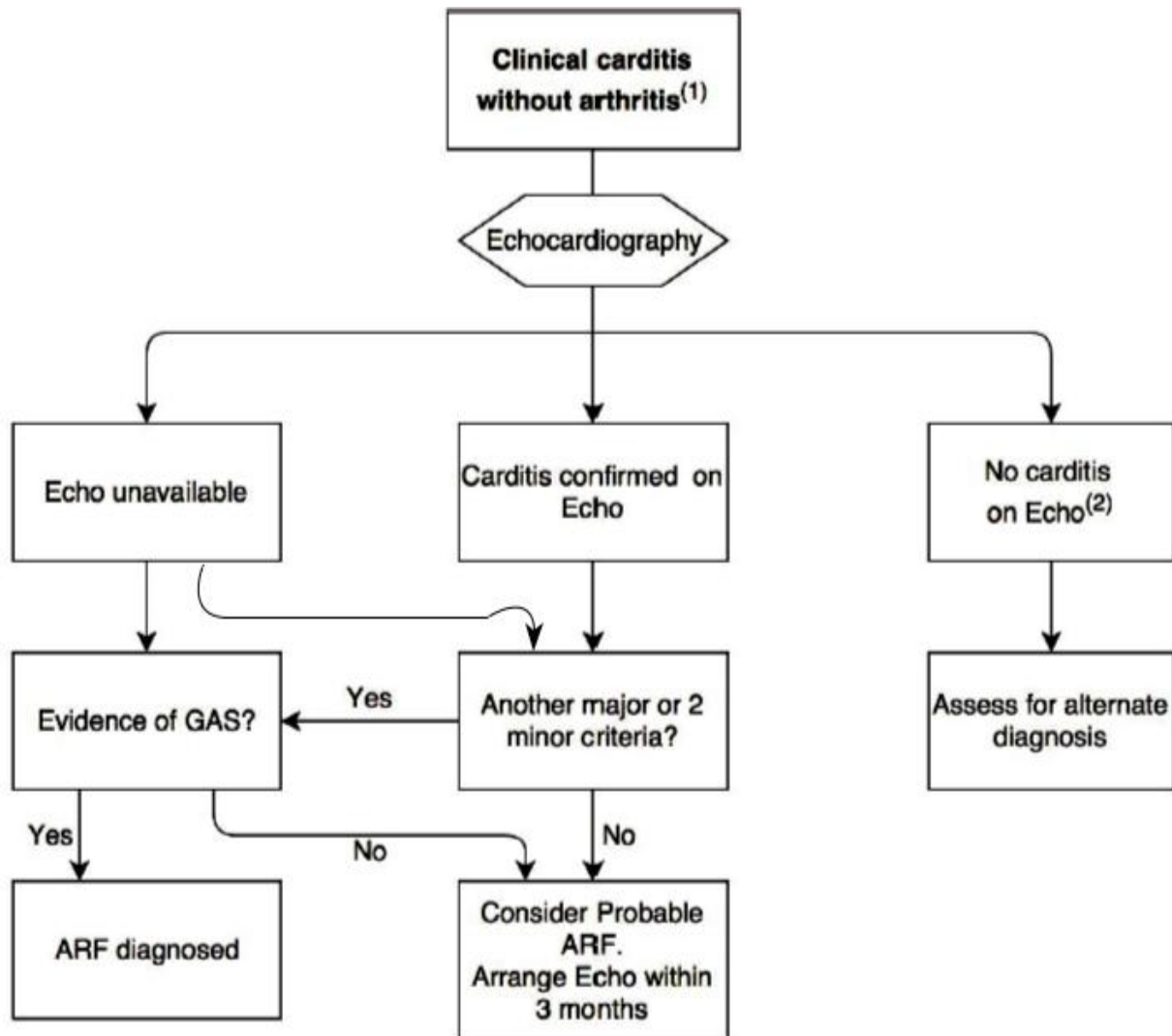
## Algorithm 1: Diagnosis of ARF in patients presenting with joint pain



### Carditis

- Cardiac inflammation (carditis) in ARF almost always affects the valves (valvulitis), especially the mitral and aortic valves; although pericarditis and myocarditis may also occur.
- Valvulitis observed clinically as an apical holosystolic murmur (mitral regurgitation) with or without mid-diastolic flow murmur (mitral stenosis) or an early diastolic murmur at the base (2nd or 3rd left intercostal space) of the heart (aortic regurgitation).[25]
- Early disease leads to valvular regurgitation, whereas prolonged or recurrent disease may lead to increased valvular regurgitation with impaired ventricular function or mitral stenosis.
- The rheumatic aetiology can usually be confirmed on echocardiography (see Appendix 1)
- Echocardiographic evidence of sub-clinical carditis can also be accepted as a major criteria (Table 2).
- The incidence of carditis in the initial attack of ARF varies between 50% and 82%. [26]
- Diagnosis of ARF in patients presenting with clinical carditis shown in Algorithm 2.

## Algorithm 2: Diagnosis of ARF in patients presenting with clinical carditis, without arthritis<sup>(1)</sup>



1. For patients with arthritis, use arthristis algorithm instead

2. Echocardiography is a higher level of evidence than clinical assessment

## Sydenham's chorea

This manifestation predominantly affects females, particularly in adolescence. [27, 28]

Chorea consists of jerky, uncoordinated movements, especially affecting the hands, feet, tongue and face. The movements disappear during sleep. They may affect one side only (hemichorea) or both sides of the body.

Useful signs include:

- The 'milkmaid's grip' (rhythmic squeezing when the patient grasps the examiner's fingers)
- 'Spoonning' (flexion of the wrists and extension of the fingers when the hands are extended)
- the 'pronator sign' (turning outwards of the arms and palms when held above the head)
- Inability to maintain protrusion of the tongue.

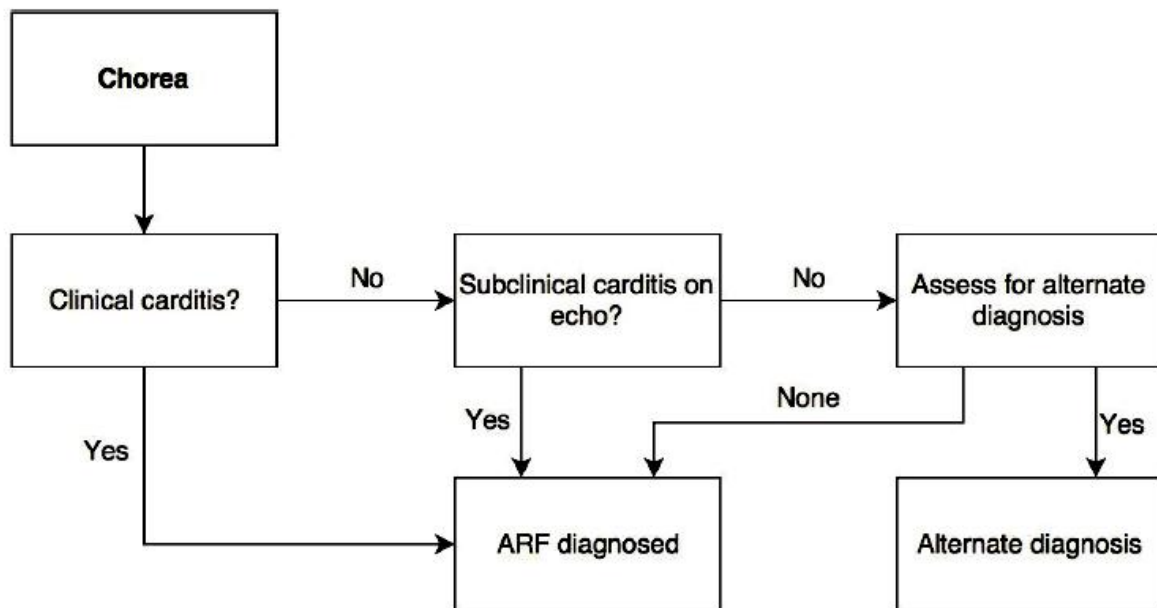
Because chorea may occur after a prolonged latent period following GAS infection, the diagnosis of ARF under these conditions does not require the presence of other manifestations or evidence of a recent GAS infection. Patients with pure chorea may have a mildly elevated ESR (approximately 40 mm/h), but have a normal serum CRP level and white cell count.

Echocardiography is important for cases of chorea without clinical carditis. Most studies show > 50% of cases of chorea have subclinical carditis, which makes the diagnosis of ARF certain. If echo is normal, it is necessary to investigate for the differential diagnoses of chorea (Table 4). Specialist paediatrician or physician involvement is recommended.

Chorea is the ARF manifestation most likely to recur, and may be associated with pregnancy or oral contraceptive use. The vast majority of cases resolve within 6 months (usually within 6 weeks), although rare cases lasting as long as 3 years have been documented. Chorea patients have a higher-than-expected prevalence of attention-deficit hyperactivity disorder, anxiety, depression and cognitive dysfunction after they have recovered from the movement disorder, although there is some evidence that attention-deficit hyperactivity disorder and anxiety features are often present before the onset of chorea, suggesting that they may be risk factors, rather than long-term complications.[29-32]



### Algorithm 3 : Diagnosis of ARF in patients presenting with chorea



### Subcutaneous Nodules

- These are very rare (less than 2% of cases), but highly specific manifestations of ARF.
- They are 0.5–2 cm in diameter, round, firm, freely-mobile and painless nodules that occur in crops of up to 12 over the elbows, wrists, knees, ankles, Achilles tendon, occiput and posterior spinal processes of the vertebrae.
- They tend to appear 1–2 weeks after the onset of other symptoms, last only 1 - 2 weeks (rarely more than 1 month); they are strongly associated with carditis.

### Erythema Marginatum

- Erythema marginatum is also rare, being reported in less than 2% of cases in Aboriginal people and populations of developing countries.
- As with subcutaneous nodules, erythema marginatum is highly specific for ARF
- It occurs as bright pink macules or papules that blanch under pressure and spread outwards in a circular or serpiginous pattern. The rash can be difficult to detect in dark-skinned people, so close inspection is required. The lesions are not itchy or painful, and occur on the trunk and proximal extremities and almost never on the face.
- The rash is not affected by anti-inflammatory medication, and may recur for weeks or months, despite resolution of the other features of ARF.
- The rash may be more apparent after showering.

## Differential diagnosis of common major manifestations

It is important to consider differential diagnosis when considering each of the major criteria of ARF. The Jones Criteria update published in 2015, likely influenced by the New Zealand and Australian guidelines, provides a list of alternate diagnoses to consider in the evaluation of patients with arthritis, carditis or chorea. This list has been further modified for Fiji to include high incidence infectious diseases such as Dengue Fever, Chikungunya and Zika Virus (See Table 4).

**Table 4: Differential Diagnoses of Arthritis, Carditis and Chorea to be considered in Fiji**

Arthritis	Carditis	Chorea
Septic arthritis	Physiological mitral regurgitation	Drug intoxication
Arboviruses including Dengue, Chikungunya, Zika Fever	Mitral valve prolapse	Wilson disease
Other viral arthritides (e.g. parvovirus)	Congenital mitral valve disease	Encephalitis
Reactive arthritis	Congenital aortic valve disease	Familial chorea
Endocarditis	Myocarditis	Tic disorder
Typhoid Fever	Cardiomyopathy	Cerebral palsy
Juvenile Idiopathic Arthritis	Kawasaki disease	SLE
SLE and other auto-immune disease		

SLE: Systemic lupus erythematosus

### 1.4.2 Minor Manifestations

#### Monoarthralgia

- Monoarthralgia is a highly non-specific clinical presentation, but may form part of the presentation of ARF.
- Alternative diagnoses should be considered in a patient with arthralgia involving a single joint that is not typical of ARF.

#### Fever

- Most manifestations of ARF are accompanied by a fever (with the exception of chorea).
- On oral, tympanic or rectal temperature  $\geq 38^{\circ}$  C on admission or documented during the current illness.

## Elevated acute phase reactants (ESR or WCC)

- An ESR of  $\geq 30$  mm/h meets the diagnostic criterion
- ESR in ARF is typically  $> 80$ mm/h and usually remains elevated for  $> 4$  weeks, and may remain elevated for 3 – 6 months following an episode of ARF.
- A white cell count  $\geq 15 \times 10^6/\text{mL}$
- C reactive protein is included in the 2015 Jones criteria rather than WCC
- WCC is included in the WHO 2003 criteria for ARF
- CRP is not available in Fiji, therefore WCC has been included.

## Prolonged PR interval

- An ECG should be performed in all cases of suspected ARF
- The PR interval normally increases with age and therefore needs to be age-adjusted (Table 5)

**Table 5 : Upper limits of normal P-R interval**

Age group (years)	Seconds
3-12	0.16
12-16	0.18
$>17$	0.20

- Prolonged PR is occasionally a normal variant, but one that resolves over days to weeks may be a useful diagnostic feature of ARF when other clinical features are not definitive.
- If a prolonged PR interval is detected, the ECG should be repeated after two months.
- Advanced AV block occurs in 5% of patients (i.e. 2nd and 3rd degree block, and tachy-arrhythmias.[25]

## Echocardiography and diagnosis of ARF

Echocardiography for acute carditis is more sensitive and specific than auscultation and it is recommended that all patients with suspected or definite ARF undergo echocardiography.[11, 33, 34] ARF and RHD auscultation-based diagnosis is now considered inadequate in the face of international evidence.[11, 35-38]

Echocardiography for diagnosis of ARF is highly recommended; even when diagnosis based on clinical signs appears certain. Echocardiography should be undertaken within 3 months of clinical diagnosis. The changes of ARF on echo are still expected to be seen within 3 months. The uses of echocardiography for ARF diagnosis are presented in Table 6 and Table 8.

**Table 6: Summary of uses of echocardiography in ARF diagnosis**

<b>Diagnosis</b>
Echocardiographic evidence of sub-clinical carditis is sufficient as a major manifestation of ARF
<b>Pericarditis</b>
Confirming the presence of a pericardial effusion Revealing inaudible or subclinical valvular regurgitation in the presence of a friction rub
<b>Myocarditis and congestive heart failure</b>
Assess left ventricular function Assessing the severity of valvulitis (valvulitis is usually present in ARF with heart failure)
<b>Valvulitis</b>
Visualisation of the anatomy of the valves, especially in mitral regurgitation (very important for surgical decision making) Defining the severity of mitral and aortic and/or tricuspid regurgitation Defining the severity of mixed valve disease Identifying subclinical evidence of rheumatic valve damage

Echocardiography is helpful in assisting clinicians where the diagnosis and clinical utility of sub-clinical carditis in managing ARF is otherwise uncertain. (Table 7) The minimal findings of ARF are shown in Table 8.

**Table 7: Diagnostic and clinical utility of sub-clinical carditis in managing ARF**

Main Clinical Features of ARF	Implications of a Finding of Sub clinical Valve Damage
	<b>Diagnostic</b>
Polyarthritis	Usually none, as Jones criteria fulfilled, but can increase confidence in diagnosis of ARF. May confirm the diagnosis
Monoarthritis or arthralgia	ARF, as long as other causes of joint disease are excluded. Confirms the diagnosis as ARF
Chorea	Avoids the need to exclude other causes of chorea
Erthema marginatum	Nil, because clinical carditis or polyarthritis usually present
Subcutaneous nodules	Nil, because clinical carditis or polyarthritis usually present
Clinical carditis	Confirm or returns clinical diagnosis of carditis
	Helps to reinforce the importance of secondary prophylaxis
	Defines involvement of second valve if one 1 valve has clinical carditis

**Table 8: Minimal echocardiographic diagnostic criteria for the diagnosis of pathological valvular regurgitation**

<b>Mitral Regurgitation</b>
<ul style="list-style-type: none"> <li>- Seen in two views</li> <li>- In at least one view jet length &gt; 2cm</li> <li>- Peak velocity <math>\geq 3\text{m/ second}</math></li> <li>- Pan-systolic jet in at least one envelope</li> </ul>
<b>Aortic Regurgitation</b>
<ul style="list-style-type: none"> <li>- Seen in two views</li> <li>- In at least one view jet length &gt; 1cm</li> <li>- Peak velocity <math>\geq 3\text{m/ second}</math></li> <li>- Pan-diastolic jet in at least one envelope</li> </ul>

### **1.4.3 Management of ARF**

#### **Guidelines for general in-hospital care**

Persons with symptoms of ARF should be hospitalised to ensure accurate diagnosis, and to receive clinical care and education about preventing further episodes of ARF.

Clinicians should appreciate that the diagnosis may not be certain for a number of days as:

- (a) arthritis may evolve
- (b) delay in test results and
- (c) need for repeat tests such as ASOT.

The diagnosis should include an initial echocardiogram (if available) to help identify and measure heart valve damage. Long-term preventative management should be organized before discharge.

## Box 4: Treatment of Acute Rheumatic Fever

### The symptoms of acute rheumatic fever should be managed as follows:

Paracetamol and/or codeine should be used to control arthritis, fever, and other acute symptoms **until** the diagnosis of ARF is confirmed. Once confirmed, high dose salicylates (aspirin) are the preferred choice, although other NSAIDs (e.g. ibuprofen) may be equally effective. Steroids may rarely be needed. Use:  
Aspirin 60–100 mg/kg/day, orally, given in 4 divided doses (maximum of 8 g/day) for 1–2 weeks, then wean according to clinical response and inflammatory markers (CRP, ESR): usual duration 6–8 weeks but longer duration may be required.

Carditis resulting in heart failure is treated with standard therapies (diuretics, ACEI), and cardiac arrhythmias that may develop are treated accordingly (see Section 11 Acute management of arrhythmias).

Steroids have been used, but conclusive evidence of their efficacy is limited. Where a decision is made to use steroids in severe carditis, use:

Prednisolone 1–2 mg/kg/day to a maximum dose of 80 mg daily: therapy is usually continued for 1–3 weeks; where >1 week is required, wean by 20–25% each week.

In severe disease, bed rest is recommended.

Chorea is usually managed conservatively, however carbamazepine and sodium valproate may be used for control.

ARF is a notifiable disease. Prior to discharge, complete the Notifiable Disease Form and dispatch it to the Health Information Unit.

Further, ensure that all items on the ARF/RHD pre-discharge form are completed and that notification form is sent to the National RHD Prevention and Control program for inclusion in the national RHD database. Call 331 9348 or email <fijirhd@gmail.com>.

Source: Fiji Cardiovascular Therapeutic Guidelines 2015

## Box 5: Acute Clinical Management of ARF

### Nursing observations

Temperature, pulse, respiratory rate, blood pressure 4 times daily Sleeping pulse (e.g. 0200 hours)

If pulse >100, apical heart rate

### Diet

Free fluids (if no heart failure)

Normal diet (limit extras)

Early diet advice, if overweight and in failure, to avoid further weight gain Weekly weight

### Bed rest and general care

Strict bed rest not necessary for most patients Plan care to provide rest periods

Provide age-appropriate activities

Notify school teacher

Involve family in care

Prepare for discharge to primary care facility, and follow up

### If clinical carditis present (heart murmur, heart failure, pericardial effusion, valvular damage)

Document cardiac symptoms and signs

Daily weight and fluid balance chart

Diuretics, ACE inhibitors, digoxin, if indicated; consider glucocorticoids Anti-coagulation if atrial fibrillation present

Cardiology opinion

ACE: angiotensin-converting enzyme, IM: intramuscular, IV: intravenous.

## **Notification**

ARF was made a nationally notifiable disease to National Infectious Disease Surveillance team in 2006. The MoHMS National Acute Rheumatic Fever and Rheumatic Heart Disease Policy 2015 states in sections:

- All incident cases of ARF/RHD will be reported through the Notification of Notifiable Disease and a registration form for ARF/RHD will be completed and send to the Divisional RHD coordinator, and
- All cases of ARF/RHD will be flagged on the Patient Information System (PATIS) for general information.

## **Secondary prophylaxis of ARF**

Secondary prophylaxis is the term used to describe regular delivery of antibiotics to prevent recurrence of Group A Streptococcal infection and subsequent development of ARF. Secondary prophylaxis is recommended for all people who have a history of ARF or RHD. The most effective method of secondary prophylaxis is Benzathine penicillin G given by intramuscular injection every 3 or 4 weeks. Oral Penicillin may be used but the patient and family must be aware this treatment is associated with a higher recurrence rate of ARF compared to using benzathine penicillin, partly because adhering to twice-daily tablets over many years is difficult.

Oral Erythromycin is used if there is an allergy to Penicillin.

## **Regular secondary prophylaxis:**

- Prevents the occurrence of GAS infections which can lead to recurrent ARF
- Reduces the severity of RHD (and can result in cure of RHD after many years)
- Helps prevent death from severe RHD.

## **Indications for Use**

Secondary prophylaxis is indicated for people who have

- ARF confirmed by the Jones Criteria
- RHD confirmed on echocardiogram
- ARF or RHD not confirmed, but highly suspected.



## **Adherence to secondary prophylaxis**

A variety of factors combine to limit the uptake of long-term secondary prophylaxis. Primary care facilities should be aware of any local barriers to receiving secondary prophylaxis, and work within the system and with patients and families to reduce these barriers. For example, adherence has been seen to improve when patients feel a sense of personalised care and 'belonging' to the clinic, and when recall systems extend beyond the boundaries of the community. [39]

Hospitalisation at diagnosis provides an ideal opportunity to begin or re-establish secondary prophylaxis, and to educate patients and families on how important it is to prevent future episodes of ARF. Appropriate continuing education and support by primary care staff should continue once the patient has returned home. A recent study undertaken in Fiji has highlighted the importance of a supportive relationship from primary health providers, particularly nurses, was key to improving patients levels of adherence to BPG. Secondary prevention of further episodes of ARF is a priority. It should include strategies aimed at improving the delivery of secondary prophylaxis and patient care, the provision of education, coordinating available health services and advocacy for necessary and appropriate resources.

### **Strategies to promote continuing adherence include:**

- Identifying local, dedicated public health nurses to support delivery of secondary prophylaxis and coordinate routine care within medical area.
- Focusing on improving relationships between health staff and patients/families.
- Developing and implementing recall and reminder systems (based on a local ARF/RHD register where established) to accommodate the high mobility of individuals and groups.
- Ensuring that recall systems extend beyond community/medical areas.
- Establishing networks for timely communication between health clinics.
- Using a centralised coordinator and register to assist in monitoring movement.
- Minimising the impact of staff turnover where possible.

- Educating about the importance of secondary prophylaxis in preventing recurrent ARF and the development or worsening of RHD.
- Improving the quality and delivery of ongoing health education and support for staff, patients and families.
- Implementing measures to reduce pain of injections.
- Basing routine care on standardised evidence-based guidelines.
- Engaging school health teams to provide support and counselling to patients and raise awareness among teaching personnel about importance of secondary prophylaxis.

## Duration of Secondary Prophylaxis

Each individual who requires secondary prophylaxis requires independent evaluation as there are a number of additional factors that can help determine the length of time required for treatment. The following points should be considered carefully when planning the duration of secondary prophylaxis.

1. **Age.** ARF recurrence is less common after age 25 and rare after age 40, therefore younger people may need treatment for longer.
2. **Severity of RHD.** An additional ARF illness could be life-threatening for people with moderate or severe RHD and following valve surgery, therefore people with more complex disease may need to remain on treatment for longer.
3. **Carditis during initial ARF.** Early heart damage increases the risk of further damage with recurrent ARF.
4. **Length of time since last ARF.** ARF recurrence is less common more than 5 years since last episode.
5. **Medication delivery.** Regular prophylaxis in the first few years after the initial ARF may provide greater protection from recurrences than irregular prophylaxis for many years.
6. **Disease progression.** Evidence of worsening RHD at any stage require extended prophylaxis.

Recommended duration for secondary prophylaxis regimens are included in Table 9.

**Table 9: Recommended secondary prophylaxis regimens**

Category	Definition of Category	Duration
Minimum duration for all persons with ARF or RHD <sup>1</sup>		Minimum 10 years after most recent episode of ARF or until age 21 years (Whichever is longer)
No RHD	No pathological mitral or aortic regurgitation, but may have minor changes to mitral or aortic valves on Echocardiography	Discontinue at that time <sup>^</sup>
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 after the minimum duration
Moderate RHD	<ul style="list-style-type: none"> <li>Any valve lesion of moderate severity clinically (e.g. mid-moderate cardiomegaly and/or mild-moderate heart failure) or on echocardiography</li> <li>Mild mitral regurgitation together with mild aortic regurgitation clinically or on echocardiography</li> <li>Mild or moderate mitral or aortic stenosis</li> <li>Any pulmonary or tricuspid valve lesion co-existing with a left-sided valve lesion</li> </ul>	Continue until 35 years of age
Severe RHD	<ul style="list-style-type: none"> <li>Any severe valve lesion clinically (e.g. moderate to severe cardiomegaly or heart failure) or on echocardiography</li> <li>Any impending or previous cardiac valve surgery for RHD</li> </ul>	Continue until age 40 years, or longer *

<sup>1</sup> Patients >25 years of age who are diagnosed with RHD, without any documented history of prior ARF, should receive prophylaxis until the age of 35 years. At this time, they should be reassessed to determine whether prophylaxis should be continued.

<sup>^</sup> Decisions to cease secondary prophylaxis should be based on clinical and echocardiographic assessment.

\* Risk of recurrence is extremely low in people aged >40 years. In some cases, for example, when the patient decides that they want to reduce even a minimal risk of recurrence, prophylaxis may be continued beyond the age of 40 years, or even for life. Continue prophylaxis for life (benzathine penicillin where not allergic) after valve replacement surgery.

The expected date to cease secondary prophylaxis should be recorded by the clinician and provided to the RHD register. Each individual should be carefully reviewed to determine any ongoing risk of ARF recurrence and the level of heart valve damage before ceasing secondary prophylaxis. **Ceasing secondary prophylaxis should only be done by a Specialist Medical Officer.**

An assessment prior to ceasing secondary prophylaxis should include

- Estimated time since last ARF illness (i.e. more than 5 years since last ARF)
- Specialist clinical review by a Medical Specialist or Cardiologist
- Echocardiogram to establish presence and severity of RHD (if available).

## **Secondary prophylaxis and pregnant women**

Penicillin and erythromycin are considered safe for use in pregnancy.

Low dose lignocaine is safe in pregnancy.[40] A large number of pregnant women and women of child bearing age have been exposed to lignocaine. Lignocaine crosses the placenta but there is no evidence of an association with fetal malformations, cardiac rhythm disturbances or other significant side effects in pregnant women or their babies.

## **Secondary Prophylaxis While Breastfeeding**

Penicillin is excreted into breast milk in low concentrations and is considered safe for use in breast feeding. Erythromycin is also excreted into breast milk and has been considered safe in breastfeeding. Monitor infants for vomiting, diarrhea and rash while breast feeding mothers are on antibiotic courses.

Lignocaine can be administered to breastfeeding women. Lignocaine is excreted into breast milk in small amounts, however the oral bio availability of lignocaine is very low (35%).[41] Given the small amount of lignocaine used with benzathine penicillin, the amount excreted into breast milk to which the infant is therefore exposed, is minimal. Lignocaine is unlikely to cause adverse effects in breast feeding infants.

## **Management plan when the ARF episode is controlled**

- Administer the first dose of secondary prophylaxis (refer to Table 10 for recommended dose and frequency)
- Notify the RHD Divisional coordinator and register the individual with the RHD Programme:
- Provide disease education for the person with ARF and the family
  - Understanding of ARF and RHD and risks of ARF recurrence
  - Importance of regular secondary prophylaxis and medical review
  - Recognising own signs and symptoms of ARF and RHD
  - Risks associated with future RHD (e.g. pregnancy, surgery and high level of aftercare)
  - Importance of dental health, and informing the dentist of a history of ARF and the need for antibiotic prophylaxis prior to a dental procedure
  - Importance of presenting to a doctor or clinic if they have a sore throat – all sore throats should be treated with an antibiotic effective against group A streptococci but which is not penicillin ie. Bactrim
  - Include an ARF diagnosis alert on computer systems and/or medical files
  - Refer to local Medical Officer / health facility for ongoing management
  - Organise future specialist reviews
  - Arrange dental review (and provide advice about endocarditis prevention)
  - Influenza vaccine (if available).

## **Long-term Management of ARF**

- Regular secondary prophylaxis
- Regular 6-12 monthly medical review
- Regular annual dental review

## **Echocardiogram following each episode of ARF, and routine echocardiogram:**

- every 2 years for children (sooner if there is evidence of cardiac symptoms)
- every 5 years for adults (sooner if there is evidence of cardiac symptoms or there is rheumatic heart disease).

## **Role of RHD divisional nurse coordinators in monitoring and reporting secondary prophylaxis**

- Work closely with health facility staff to support and monitor the delivery of Benzathine treatment to known ARF/RHD patients in assigned Medical Area
- Coordinate activities around RHD control in Fiji at a Divisional level, liaising with collaborators and stakeholders;
- Support public health staff to deliver, monitor and report care for people with ARF and RHD at primary and secondary health care level at divisional and national level;
- Oversee case finding activities and defaulter tracing, RHD data management and reporting and provide regular program reports to the National RHD Advisory Group (TAC) and Fiji RHD project team members;
- Raise awareness of RHD among health staff and the wider community in Fiji
- Coordinate and deliver regular workshops/one on one training sessions on RHD prevention and control with health workers and community;
- Attend and contribute to relevant Divisional, National and Local meetings as required

## ***Role of RHD liaison nurses***

- Work closely with health centre and zone nursing staff to support and monitor the delivery of Benzathine treatment to known ARF/RHD patients in assigned medical area
- Facilitate collection of quarterly RHD injection data within medical area
- Facilitate follow-up of Benzathine defaulters
- Facilitate communication of any permanent patient movement between facilities
- Provide quarterly Benzathine updates on all registered patients in the health facility or medical area to line manager and the Divisional RHD coordinator
- Deliver awareness sessions to health personnel in the health facility or medical area
- Work with health centre and zone nursing staff to ensure education and support is provided to patients/families

### **1.4.4 Protocol for Secondary Prophylaxis Delivery**

Benzathine penicillin G is most effectively given as a deep intra muscular injection, into the upper outer quadrant of the buttock or the antero-lateral thigh. Volumes of >2mls should not be given into deltoid muscle.

#### **Assessment and Preparation**

- Confirm the person's identity
- Review known drug allergies
- Discuss and record any ARF or RHD symptoms since last injection (refer to Medical Specialist if necessary)
- Obtain verbal consent for injection from patient and or parent/guardian.

#### **Injection Procedure**

A new, disposable syringe and needle must be used for each injection.

Benzathine penicillin G should be given immediately following preparation:

1. Check medication name and expiry date
2. Use a size 23 gauge needle for injection
3. Use water for injections as diluent and add 1ml 1% lignocaine to syringe after drawing up diluted BPG.
4. Dispose of used needles and syringes in a puncture-proof container (do not recap needle).

#### **Prepare Benzathine penicillin G solution as directed by the product information:**

- a. Administer 1,200,000 units for all persons  $\geq 20\text{kg}$
- b. Administer 600,000 units for children  $< 20\text{kg}$

## Documentation

- The following should be recorded in the Benzathine Penicillin injection book and medical record
  - Drug name, dose and batch number
  - Date injection given and date next due
  - Signature (of person giving the injection)
- Record the next date due on the individuals 's clinic reminder card
- Provide the above information to the local RHD register/Programme

## Pain Reduction

The following strategies may help to reduce pain following injection:

- Addition of Lignocaine 1% to diluent when preparing syringe
- Warm syringe to room temperature between hands
- Apply gentle pressure to the injection site for 10 seconds with the finger or thumb before injection
- Ensure that skin swabbed with alcohol is dry before injecting
- Deliver the injection slowly (preferably over 2 or 3 minutes)
- Use distraction to focus attention away from the injection
- Encourage movement (e.g. walking) following injection
- Develop a good rapport with the patient, assisted by having dedicated injection nurses who are open and friendly.

**Table 10: Antibiotic regimens for secondary prophylaxis of ARF/ RHD**

Antibiotic	Dose	Route	Frequency
<i>First line</i>			
BPG	900 mg (1,200,000 U) ≥20 kg 450mg (600,000 U) < 20 kg	Intramuscular injection	4 weekly *
<i>Second line (If im route is not possible or refused, adherence should be carefully monitored)</i>			
Phenoxymethylpenicillin (Penicillin V)	250mg	Oral	Twice daily
<i>Following documented penicillin allergy</i>			
Erythromycin	250mg	Oral	Twice daily

\* Three-weekly BPG may be considered for patients with moderate or severe carditis or a history of valve surgery, who demonstrate good adherence to less frequent injections, and for those who have confirmed breakthrough ARF, despite full adherence to 4-weekly BPG. IM: intra muscular



## Anaphylaxis

Anaphylaxis to BPG is rare. The signs and symptoms of an anaphylactic reaction include: rapid weak pulse, wheeze, tightness in the chest, pruritis (itching), urticaria (rash), dizziness or headache, flushing and or periorbital oedema.

### Response procedure:

- Do not leave the patient alone
- Call for assistance
- Lie the patient in recovery position (if severe respiratory distress may be better sitting up)
- Ensure the airway is clear, give oxygen if available
- Call for support
- Give adrenaline (Table 11)
- Check vital signs, note colour, tone and perfusion
- If further signs of deterioration, repeat adrenaline after 10 minutes
- Up to 3 doses of adrenaline can be given.

**Table 11: Adrenaline dosage for anaphylaxis**

12 years of age and older	
0.5ml of 1:1000 adrenaline, deep IM injection	
Under 12 years of age	
approximately 0.01 ml/kg of 1:1000 adrenaline, deep IM injection	
age 0-3 years	0.1 ml
age 4-6 years	0.2ml
age 6-8 years	0.3ml
Age 9-12 years	0.4ml

## **Discharge from hospital**

The duration of hospital stay is dictated by the clinical response and improvement of inflammatory markers (ESR). Most cases without severe carditis can be discharged after approximately 2 weeks. The length of admission may also depend on the social and home circumstances. If individuals come from remote or island communities with limited access to high level medical care, it may be advisable to prolong the stay in hospital until recovery is well advanced. A minimum hospital stay of 2 weeks for new ARF cases allows time for patient and family education and counselling on the importance of optimal adherence to secondary prophylaxis and regular clinical review.

- All cases and their families should have a good understanding of the cause of ARF and symptoms of an ARF episode, including early treatment of sore throats and clinical review of suspected episodes in other household or family members
- The first dose of BPG is given in the hospital setting; the patient will have subsequent doses at the local clinic or health centre
- Organisation of ongoing secondary prophylaxis delivery and communication of patient diagnosis should be facilitated by the Divisional RHD coordinator liaising with the local clinic. A discharge summary must be provided to the clinic as well as the BPG injection book and instructions for the patient to attend his/her next clinic.

## **Dental referral**

Dental care is important for people with RHD because the bacteria most likely to cause heart valve infections (infective endocarditis) often come from the mouth. Ensuring that people with RHD have access to dental care (fillings and extractions) and to dental hygiene equipment (toothbrushes and toothpaste) is an important way of minimising complications of RHD. Although there is a direct link between good dental care and improved RHD outcomes, there have been few attempts to integrate these services.[42]

Oral health for children has improved dramatically over the past 10 years, however dental care for people aged 15 years and over is not free and this impacts on frequency of review and accessibility to care. The burden of dental disease in the Pacific Islands is high, with a very small number of dentists available to serve a large population. Ensuring that people living with RHD are prioritised for dental care is an appropriate way to deflect their higher risk of complications from dental disease.

### **Reproductive health referral**

Individual counselling is the mainstay in deciding the appropriate management and reproductive health recommendations for patients with valvular disease. All patients need a family conference. A clear plan for the management of labour and puerperium in women with valvular heart disease should be established in advance, well documented and distributed widely (including to the woman herself) so that all personnel likely to be involved in the woman's intra-and post-partum care are fully informed. Further supportive counselling may be arranged with Empower Fiji, if deemed necessary by the treating physician. (For further information please refer to Fiji MoHMS Obstetrics and Gynaecology Guidelines. Version 2.1, June 2015. Chapter 5: Management of cardiac disease in Pregnancy page 59)

## **Prevention of Infective Endocarditis**

Infective Endocarditis is a serious complication of RHD and may also occur after heart valve surgery. Endocarditis is caused by bacteria in the bloodstream. This uncommonly occurs during dental or surgical procedures but often the source of the infection is not clear. In people with rheumatic valve disease, endocarditis most commonly occurs in the mitral or aortic valves since these are the most commonly damaged heart valves. Although the effectiveness of prophylactic antibiotics before dental or surgical procedures has not been proven to reduce the likelihood of developing endocarditis, they have been traditionally given as a preventive measure.

Therefore people with RHD or artificial heart valves should receive antibiotics before procedures that may introduce bacteria into the bloodstream. (see Table 12)

- People with a history of ARF but no valve damage do not require antibiotic prophylaxis before procedures.
- People who receive regular BPG or oral Penicillin for secondary prophylaxis should be offered a different antibiotic to prevent endocarditis (see Table 13)
- All people with ARF and RHD should have regular dental care to prevent dental decay and the potential risk of endocarditis

**Table 12: Procedures for which Endocarditis prevention is recommended**

DENTAL PROCEDURES	OTHER PROCEDURES
Dental extractions	Tonsillectomy/adenoidectomy
Periodontal procedures	Bronchoscopy with a rigid bronchoscope
Dental implant placement	Surgery involving the bronchial mucosa
Gingival surgery	Sclerotherapy of oesophageal varices
Initial placement of orthodontic appliances	Dilatation of oesophageal stricture
Surgical drainage of dental abscess	Surgery of the intestinal mucosa or biliary tract ( <i>except for endoscopy, biopsy and percutaneous endoscopic gastrostomy</i> )
Maxillary or mandibular osteotomies	Endoscopic retrograde cholangiography
Surgical repair or fixation of a fractured jaw	Prostate surgery
Endodontic surgery and instrumentation	Cystoscopy and urethral dilatation
Intra-ligamentary local anaesthetic injections	Vaginal delivery in the presence of infection, prolonged labour or prolonged rupture of membranes
Dental cleaning where bleeding is expected	Surgical procedures of the genitourinary tract in the presence of infection ( <i>e.g. urethral catheterisation, uterine dilatation and curettage, abortion, sterilisation, placement or removal of intrauterine contraceptive devices</i> )
Placement of orthodontic bands	

**Table13: Suggested prophylactic antibiotic regimens for dental, oral, respiratory tract and other surgical procedures**

Low risk patients (i.e Without prosthetic valves or previous history of endocarditis) undergoing dental treatment, oral surgery or procedures of the respiratory tract.
<p>Azithromycin</p> <p>Adult dose: 500mg PO</p> <p>Paediatric dose: 15mg/kg PO; not to exceed 500mg/dose</p>
Low risk patients having gastrointestinal or urinary procedures or other major surgery, and HIGH risk patients (eg. Prosthetic valve, previous history of endocarditis) undergoing and type of dental or surgical procedures.
<p>Vancomycin</p> <p>20mg/kg IV (max 500gm - infused over 1 hour) started 60 minutes before procedure. May repeat after 12 hours</p>

## **Section 2: Guidelines and clinical pathways for Rheumatic Heart Disease**

### **Introduction**

Rheumatic heart disease is the result of damage to the heart valves that occurs after repeated episodes of ARF. Early diagnosis of RHD is very important so that secondary prophylaxis can be started as soon as possible to help prevent the progression of the valve disease (see section on Secondary Prophylaxis). Echocardiography is essential to confirm the diagnosis; all patients with RHD or a past history of ARF require echocardiography. Echocardiography can grade the severity of disease and assess left ventricular size and systolic function. Echocardiography excludes false positive clinical diagnoses of RHD saving the patient unnecessary penicillin injections. The mitral valve is affected in over 90% of cases of rheumatic heart disease. The next most commonly affected valve is the aortic valve; usually disease of the aortic valve is associated with concurrent disease of the mitral valve. The tricuspid and pulmonary valves can be affected but RHD should not be diagnosed as the cause in the absence of mitral or aortic RHD. Tricuspid regurgitation (TR) may occur in advanced mitral valve disease secondary to pulmonary hypertension and TR may be severe and contribute significantly to cardiac failure. Mitral regurgitation is the most common heart valve lesion in RHD – as an isolated lesion it is found most commonly in children and young adults. Mitral stenosis represents longer-term chronic changes to the mitral valve – it is therefore more commonly seen in adults. A common complication of mitral stenosis is atrial fibrillation. Aortic regurgitation is not uncommon but aortic stenosis is almost never seen as an isolated lesion.

### **Key elements of best practice Rheumatic Heart Disease care**

The fundamental goal for the long-term management of RHD is to prevent recurrences of ARF, which lead to the progression of valve disease.

Best practice care includes:

- Secondary prevention with penicillin prophylaxis.
- Timely review by a clinician experienced in RHD management: physician or paediatrician, infectious disease specialist or cardiologist.

- Access to echocardiography for diagnosis and to assess the left ventricular function and valve function.
- Access to monitoring of anticoagulation therapy in patients with atrial fibrillation and /or mechanical prosthetic valves.
- Access to oral health care.
- Access to reproductive health care.
- Access to cardiac surgery referral.

## 2.1 Diagnosis of RHD

### Signs and symptoms

- Heart murmurs – see physical examination section below
- Heart Failure
- The symptoms of RHD depend on the valve lesion and its severity. Symptoms of RHD may not show for many years until valve disease becomes severe. Patients can have severe valve regurgitation and be asymptomatic. Box 6 shows Initial symptoms of early heart Failure

### **Box 6: Initial symptoms of RHD are the symptoms of early heart failure**

1. Breathlessness on exertion
2. Feeling tired & General weakness  
As heart failure progresses, other symptoms may develop including:
3. Cannot lie flat in bed (called orthopnoea or breathlessness on lying down)
4. Waking at night with shortness of breath (called paroxysmal nocturnal dyspnoea)
5. Swollen ankles
6. Palpitations may occur if atrial fibrillation is present (particularly in mitral stenosis)
7. People with aortic valve disease may experience angina and syncope in addition to heart failure symptoms.

## Physical Examination

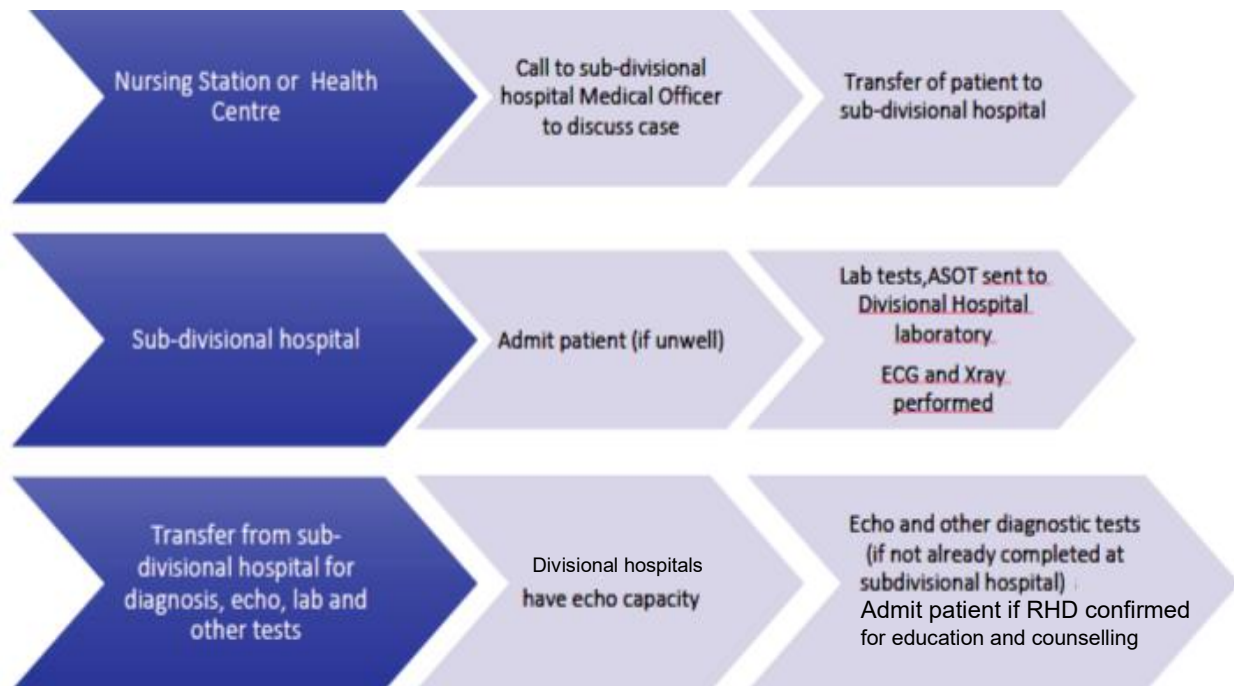
Clinical assessment should include examination of blood pressure, feeling the cardiac impulse (“precordium”) as well as listening for heart murmurs. Careful auscultation should be undertaken and suspicious heart murmurs referred for assessment by a medical specialist with echocardiography. If echocardiography is not available immediately a referral must be undertaken within 3 months of suspected RHD diagnosis. Clinical examination should include assessment of severity and complications including signs of heart failure, the presence of atrial fibrillation and any new murmurs.

- In **mitral regurgitation** the characteristic murmur is a pansystolic murmur heard loudest at the apex and radiating laterally to the axilla.
- In **mitral stenosis** the characteristic murmur is a low-pitched, diastolic rumble heard best at the apex with the bell of the stethoscope and with the person turned to their left side.
- In **aortic regurgitation** the characteristic murmur is a diastolic blowing decrescendo murmur best heard at the left sternal border with the person sitting up and leaning forward in full expiration. The BP shows a wide pulse pressure confirming the sign of collapsing pulses.
- In **aortic stenosis** (uncommon in RHD) the characteristic murmur is a loud, low-pitched mid-systolic ejection murmur best heard in the aortic area, radiating to the neck.

For more detailed descriptions of the signs and symptoms, evolution and optimal medical management of the categories of RHD the reader is referred to Walsh et al [43] and the Australian RHD guidelines 2012[3]



**Figure 2 : Referral pathway for RHD patients**



### **Diagnostic tests**

- All persons with murmurs suggestive of valve disease, or a past history of ARF, should have an echocardiogram. Echocardiography will detect any rheumatic valve damage and help determine its severity and assess left ventricular function. Regular echocardiography helps to detect evidence of progression of valve lesions over time and to assess heart function before surgery.
- All assessments of a new murmur or established rheumatic heart disease should include ECG and CXR. ECG is essential to determine rhythm. The CXR helps to assess the size of the heart chambers and to detect pulmonary congestion.

Echocardiography easily shows the aetiology is rheumatic in severe RHD. In 2012 consensus guidelines were published for the minimal echocardiographic diagnosis of RHD. The guidelines were developed by a group of international experts under the auspices of the World Heart Federation (WHF) and published in 2012.[44] See Appendix 1.

## 2.2 Management of RHD

### ***Management of RHD depends on the severity of disease.***

The management of severe RHD is complex and requires careful co-ordination. The main goal is to prevent disease progression and to avoid, or at least delay, valve surgery. Secondary prophylaxis for prevention of recurrent ARF is the main strategy to achieve this (refer page 31 Protocol for Secondary Prophylaxis). Regular clinical review is essential and follow-up echocardiography is important to follow the progress of the heart valve lesions.

The management of milder RHD is based on delivery of excellent secondary prevention as it has been shown that progression to more severe RHD will not occur in the absence of recurrences of ARF. Thus nursing follow up is key, with physician review less frequently required (Table 14).

### ***Clinical management***

The fundamental goals in the long-term management of RHD are to prevent ARF recurrences, which lead to the progression of valve disease, and monitor left ventricular function. In many cases with mild or moderate RHD, resolution of RHD is observed over time. In mild and moderate RHD the left ventricle is not at risk of failing. Unfortunately, even without recurrences of ARF, those with severe RHD may develop progression of disease due to the irreversible damage of the valves and left ventricular myocardium.

All patients with RHD or a past history of ARF who develop new heart murmurs require echocardiography

- Echocardiography can grade the severity of valvular disease and assesses left ventricular (LV) size and systolic function.
- Serial echocardiographic data plays a critical role in determining the timing of any surgical intervention.

Clinicians have a key role to reinforce the need for secondary prophylaxis for their patients.

## ***Dental care***

Routine dental care is critically important in cases with a history of ARF and/or RHD. All patients should receive education about oral hygiene, and should be referred promptly for dental assessment and treatment when required. This is especially important prior to valvular surgery, when all oral/dental pathology should be investigated and treated accordingly.

- People with rheumatic heart disease have an increased risk of developing infective endocarditis, a condition with significant morbidity and mortality.
- Oral bacteria have been identified as causative agents for infective endocarditis and so it is important that all people with RHD have meticulous dental and oral hygiene to reduce the risk of oral source for bacteraemia.
- Regular oral health care, which includes assessment, treatment and preventive education should be part of the routine ongoing management of RHD. It is recommended that all patients with rheumatic heart disease (regardless of severity) undergo at least an annual oral health review.
- Dental recall intervals should be based on clinical risk, therefore people with moderate/severe RHD, prosthetic cardiac valves or higher dental risk factors (e.g. poor oral hygiene, dry mouth, untreated dental caries and inflammatory periodontal disease) should be offered more frequent dental reviews based on clinical risk.
- Some dental procedures do have an increased risk of causing an oral bacteraemia.
- Antibiotic prophylaxis is recommended for at risk patients having at risk dental procedures.

## ***Clinical review***

The frequency of clinical review should be determined according to individual needs and local capacity. Most importantly review should become more frequent in the event of new symptom onset, symptomatic deterioration or a change in clinical findings - see Table 14.

**Table 14 : Recommended routine clinical review and management plan for RHD[25]**

Classification	Criteria	Review and Management Plan				
		Secondary prophylaxis	Cardiologist, Physician or Paediatrician review	Echocardiography	Dental Review	Endocarditis Prophylaxis
Low Risk	ARF with no evidence of RHD or Trivial or mild valvular disease	4 weekly	3-5 yearly by paediatrician or physician	If new murmur or change in examination findings	Yearly	Recommend if mild valvular involvement
Medium Risk	Moderate RHD	4 weekly	2-3 yearly	2-3 yearly	Yearly	Recommended
High Risk**	Any severe valve lesion or Mechanical prosthetic valves or Tissue prosthetic valves or valve repair	3 or 4 weekly	6-12 monthly	6-12 monthly	6 monthly	Recommended

\* Fiji RHD programme historically recommends 3 weekly (every 21 days) Benzathine penicillin dosing for children aged < 15 years (see below section)

\*\* Anyone with severe valvular disease or moderate to severe valvular disease with symptoms should be referred for cardiology and surgical assessment as soon as is possible

## **Evidence for optimal delivery of Benzathine Penicillin G for Acute Rheumatic Fever and Rheumatic Heart Disease**

Secondary prophylaxis with BPG is recommended for all people with a history of ARF or RHD. Achieving optimal BPG adherence to provide a protection against ARF to reduce days at risk for recurrence is the mainstay of ARF/ RHD control.

In Fiji a three weekly (every 21 days) regimen has historically been in place for children aged less than 15 years; requiring a total of 17 injections to be delivered per year. Adolescents and adults are prescribed a four weekly (every 28 days) regimen; requiring a total of 13 injections per year.

International evidence shows that four-weekly BPG is currently the treatment of choice, except in patients considered to be at high risk, for whom three-weekly administration is recommended.[3, 5, 25]

- The benefits of three-weekly BPG injections are offset by the difficulties of achieving good adherence.
- Data from the Northern Territory, Australia show that few, if any, recurrences occurred among people who fully adhered to a four-weekly BPG regimen. [45]
- Data from Fiji show that many children do not achieve optimal adherence on the current three weekly regimen. [46]

## **Secondary prevention**

Refer page 31 Protocol for Secondary Prophylaxis

## **Anticoagulation therapy**

All patients with atrial fibrillation and valvular heart disease (including rheumatic heart disease) should be recommended for long-term anticoagulation where not contraindicated. (Fiji Cardiovascular guidelines 2015).[47] Warfarin, a vitamin K antagonist, is the drug of choice for anticoagulation for patients with prosthetic heart valves, for both RHD and non-rheumatic valve disease. As the absorption of warfarin is affected by diet and other medications, the international normalised ratio (INR) must be measured on a regular basis with adjustments of the dose as required (Box 7).

## **Box 7 : Recommended therapeutic INR ranges**

The usual recommended INR ranges[25] are:

- Prosthetic mitral valves 2.5 - 3.5
- Prosthetic aortic valves 2.0 - 3.0
- Both Mitral and aortic prosthetic valves 2.5 - 4.0

The clinician should specify the INR range on an individual patient basis. Titrating the warfarin dose can be difficult even with good access to INR monitoring. Inadequate INR monitoring with low levels predisposes to valve thrombosis, thromboembolism and stroke. High INRs can lead to spontaneous bleeding with a risk of stroke. The patient should be encouraged to be active in their INR control, ideally holding their own INR card, the warfarin dosage and the date of their next test. Patients on Warfarin should be monitored closely by the treating clinician; this can be challenging in Fiji with limited access to INR monitoring. (For further information regarding anticoagulation therapy, see Fiji Cardiovascular Guidelines 2015 Chapter 18: Antithrombotic Therapy page 140 and Annex F page 167)[47]

## **RHD and Pregnancy**

Pregnancy causes stress to the heart and may make any existing valve problem worse. Physiological changes that occur during pregnancy are:

- A 30-50% increase blood volume
- A reduction in systemic and pulmonary resistance
- An increased cardiac output
- An increase in heart rate by 10-15 beats per minute.

These changes will exacerbate existing valvular heart disease especially mitral stenosis and may cause life-threatening complications during pregnancy and can threaten the life of the woman and the fetus. Women with RHD are at high risk of complications. Sub-clinical RHD may be identified for the first time during pregnancy or soon after delivery when a woman develops symptoms of decompensated heart failure such as dyspnea, orthopnea and paroxysmal nocturnal dyspnea.[48]

Ideally, all women with known ARF/RHD should be fully assessed before pregnancy occurs. Women at particular high risk may be counselled to avoid pregnancy (e.g. severe pulmonary hypertension). When pregnancy occurs, management depends on the type and severity of heart valve disease. ***It is essential that a pregnant woman be assessed by a medical specialist as early as possible so that a coordinated pregnancy management and follow-up can be planned.***

Special attention should be given to women with high risk RHD including women with

- mitral stenosis (common)
- aortic stenosis (rare)
- atrial fibrillation
- prosthetic heart valves
- those receiving anticoagulant therapy with warfarin.

### **Clinical Management during Pregnancy**

Specialist management by a team of high maternal risk obstetricians, physicians and cardiologists affords optimal care for those with RHD.

- Women should have regular cardiac reviews, with the frequency determined by the severity of disease and clinical symptoms.
- Women with severe disease may require cardiac evaluation every two to four weeks after 20 weeks' gestation, especially if there is clinical deterioration.

### **Mitral/Aortic Regurgitation**

- Pregnancy is well tolerated in the majority of women with mild or moderate mitral regurgitation (MR) or aortic regurgitation (AR) especially if they are single valve lesions.
- Some women with severe MR or AR may develop congestive heart failure, especially during the third trimester and may need diuretics and vasodilator therapy.
- Angiotensin receptor antagonists and ACE inhibitors are contraindicated in pregnancy.
- Calcium channel blockers (e.g. nifedipine) or nitrates can be used in pregnancy.

## **Mitral Stenosis**

- Mitral stenosis is a common valvular lesion in RHD and can pose particularly dangerous problems during pregnancy and at delivery.
- In mitral stenosis the narrowed mitral valve limits flow across the valve during diastole which leads to reduced filling of the ventricle resulting in reduced stroke volume, cardiac output and aortic pressure.
- The tachycardia of pregnancy further shortens diastolic filling time exacerbating the impact of MS.
- The increase in blood volume during pregnancy also exacerbates the increases in left atrial pressure leading to pulmonary vascular hypertension and pulmonary oedema.
- Women with moderate or severe mitral stenosis must be cared for by a multidisciplinary team consisting of an obstetrician and cardiologist and/or obstetric physician in an appropriate clinical centre (with a neonatal team).
- Balloon mitral valvuloplasty may be indicated at the end of the second trimester for those with severe mitral stenosis (requires overseas referral).

## **Aortic Stenosis (rare)**

- Mild aortic stenosis is usually well tolerated.
- Women with moderate or severe AS, although much less common, are at significant risk of adverse maternal and fetal outcomes.
- Myocardial ischaemia may occur.
- Women with any degree of aortic stenosis must be cared for by a multidisciplinary team consisting of an obstetrician and cardiologist and/or obstetric physician in an appropriate clinical centre (with a neonatal team).
- Beta-blockers are given for rate control.
- Diuretics are prescribed to prevent volume overload.

## **Mode of delivery**

- The mode of delivery should be determined by obstetric indications with vaginal delivery or assisted vaginal delivery (with vacuum extraction or forceps) being the goal in the majority of women who have stable cardiac symptoms.
- An early epidural may be helpful in minimising the sympathetic response to labour of tachycardia and raised blood pressure.



## **Mechanical Prosthetic Heart Valves and anticoagulation in Pregnancy**

- Women with mechanical prosthetic heart valves are a very high-risk group as they must continue therapeutic dose anticoagulation throughout pregnancy to prevent valve thrombosis and its sequelae of valve failure and systemic thromboembolism.
- All anticoagulation options carry maternal and/or fetal risks:
  - Risk of warfarin embryopathy in the first trimester
  - Risk of adverse outcome(s) with any anticoagulant approach
  - Warfarin: most effective at preventing maternal thromboembolic complications, but with a high rate of fetal complications
  - Therapeutic dose adjusted low molecular weight heparin (LMWH) is less effective at preventing maternal thromboembolic complications, but results in improved fetal outcomes. Low molecular weight heparins have more predictable anticoagulant activity and a longer half-life than unfractionated heparin (UFH). They are administered subcutaneously once or twice daily without the need for monitoring. LMWH is currently unavailable on the Fiji essential drugs list, but is available for purchase at some retail pharmacies. (For further information about LMWH see Fiji Cardiovascular guidelines page 141). LMWHs are predictable and there is no need for monitoring in most situations. Impaired kidney function is a much more important risk for bleeding with LMWH than with UFH. [25, 47, 49]
- Patients with mechanical prosthetic valves should be given appropriate contraceptive advice to avoid unplanned pregnancy, and counselled about the risks to mother and fetus of any pregnancy.[49]

## **Secondary prophylaxis in pregnancy**

As there is no evidence of teratogenicity, penicillin prophylaxis should continue throughout pregnancy for the prevention of recurrence of ARF. Erythromycin is also considered safe in pregnancy.[5]

## **Complications of RHD**

For information on prevention and management of Atrial Fibrillation, Infective endocarditis and Stroke see Fiji MoHMS Cardiovascular Guidelines (2015).

## **Surgery for rheumatic heart disease**

In general it is only those with severe valve lesions that will need cardiac surgery.

- The need for surgery is determined by the severity of symptoms, evidence that the heart valves are significantly damaged and left ventricular chamber size and function.
- Surgery to repair or replace damaged heart valves is important to prevent left ventricular dysfunction and severe pulmonary hypertension.

Recommendations for cardiac surgery for adults are abridged from the AHA/ACC guidelines based on all relevant data internationally.[50] (Table15)

## **Assessment for cardiac surgery**

Echocardiography before surgery helps to assess the severity of valve disease and left ventricular function. If echocardiography is not available, a diagnosis of valve disease must rely on careful clinical examination, ECG and chest X-ray before an individual is referred to a cardiac surgical team/centre (all suspected RHD patients must have echocardiography performed within 3 months of diagnosis). Impaired left ventricular function, atrial fibrillation, diabetes and other co-morbidities can all increase surgical risk and decrease long-term survival rates after surgery. People who require emergency surgery or re-operation have an increased morbidity and mortality following surgery. The results of surgical treatment may depend on the following:

- severity of the valve disease at the time of surgery,
- left ventricular function,
- nutritional status of the individual prior to surgery,
- long-term post-operative management (particularly anticoagulation management).

Referral consideration for cardiac surgery for adults and children are indicated in Table 15 and Table 16.

**Table 15: Indications for surgery in Adults[25]**

<b>Mitral regurgitation in Adults</b>
-Severe MR with symptoms (NYHA* class II-IV) or
-LVESD $\geq$ 40mm in adults
-Impaired LV function LVEF $<$ 60%
-Pulmonary hypertension $>$ 50mmHg
- New onset AF
<b>Aortic Regurgitation in Adults</b>
-Severe AR with symptoms NYHA class II-IV or
-Asymptomatic severe AR and one of the following:
LVESD $>$ 50 mm
Impaired LV function LVEF $<$ 50%
LVEDD $>$ 65 mm
<b>Mitral Stenosis in Adults</b>
-Severe Mitral Stenosis with symptoms (NYHA class II-IV) or
- Asymptomatic severe Mitral Stenosis and one of the following:
Paroxysmal atrial fibrillation
Mitral valve area $<$ 1.5cm
Pulmonary hypertension $>$ 50mmHg
Thromboembolism

MR Mitral Regurgitation; LVESV Left Ventricular End Systolic Volume; AF Atrial Fibrillation; AR Aortic regurgitation; NYHA New York Heart Association; LVESD Left Ventricular End Systolic Dimension; LVEDD Left Ventricular End Diastolic Dimension

**Table 16: Indications for surgery in children[25]**

<b>Mitral Regurgitation</b>
<ul style="list-style-type: none"><li>- Severe MR with symptoms of breathlessness or</li><li>- Asymptomatic MR and one of the following:<ul style="list-style-type: none"><li>• Impaired LV function &lt;60%</li><li>• LVESV z-score &gt;2</li><li>• Pulmonary hypertension 50mmHg</li></ul></li></ul>
<b>Aortic Regurgitation</b>
<ul style="list-style-type: none"><li>- Severe AR with symptoms and breathlessness or</li><li>- Asymptomatic severe AR and one of the following:<ul style="list-style-type: none"><li>• LVESV z-score &gt;4</li><li>• Impaired LV function LVEF &lt; 50%</li></ul></li></ul>
<b>Mitral Stenosis in Children</b>
<ul style="list-style-type: none"><li>- Severe Mitral Stenosis with symptoms (NYHA class II-IV) or</li><li>- Asymptomatic severe Mitral Stenosis and one of the following:<ul style="list-style-type: none"><li>• Paroxysmal atrial fibrillation</li><li>• Mitral valve area &lt; 1.5cm<sup>2</sup></li><li>• Pulmonary hypertension &gt; 50mmHg</li><li>• Thromboembolism</li></ul></li></ul>

MR Mitral Regurgitation; LV Left Ventricular; LVESV Left Ventricular End Systolic Volume; AR Aortic regurgitation; NYHA New York Heart Association;

### **Contra-indications to surgery**

There are very few absolute contra-indications to valve surgery; each person must be assessed individually to determine the risks and benefits of surgery. Factors such as poor left ventricular function with valve regurgitation, severe pulmonary hypertension may pose an unacceptable risk for cardiac surgery. Age of the person and the presence of co-morbidities also affect the risk/benefit of surgery. Young people often recover well after surgery, even from severe valve disease. Co-morbidity has a much more pronounced effect in older people.

**Table 17: Review and management plan for RHD patients**

Classification	Criteria	Review and Management Plan	Frequency
Low Risk (Priority level 3)	ARF with no evidence of RHD <i>or</i> Trivial to mild valvular disease	Secondary prophylaxis	4-weekly
		Doctor review	Yearly
		Dental review	Yearly
		Echocardiogram	Children 2-yearly Adults 2-3 yearly
Medium Risk (Priority level 2)	Any moderate valve lesion in the absence of symptoms and with normal left ventricular function <i>or</i> Mechanical prosthetic valves	Secondary prophylaxis	4-weekly
		Doctor review	6-monthly
		Influenza vaccination	Yearly
		ECG	Yearly
		Medical or Cardiac specialist review	Yearly
		Echocardiogram	Yearly
		Dental review	Yearly
		Pneumococcal vaccination	5-yearly (max 3 doses)
High Risk (Priority level 1)	Severe valvular disease, or Moderate/ severe valvular disease with symptoms, or Prosthetic valves and valve repairs	Endocarditis prevention	As required
		Warfarin + Aspirin	As prescribed
		Secondary prophylaxis	3-4 weekly
		Doctor review	3-6 monthly
		Influenza vaccination	Yearly
		ECG	Yearly
		Medical or cardiac specialist review	3-6 monthly
		Echocardiogram	3-6 monthly
		Dental review	Within 3 months and yearly thereafter

### Cardiac surgical referral in Fiji

The majority of RHD patients receiving cardiac valve surgery in Fiji are operated on by one of the two visiting cardiac surgical teams that visit annually. A small number of patients are also referred overseas to India, Australia or New Zealand for cardiac surgery (<15%). The selection of suitable patients for cardiac surgery is undertaken by Fijian clinicians developing a shortlist for referral and review by visiting international cardiology teams in the weeks prior to the surgical team visit.

## **Long-term complications**

Many of the long-term complications following valve surgery are related more to the individual and the quality and frequency of management rather than to the surgical procedure or prosthesis. Complications may include prosthetic valve thrombosis and degeneration, thromboembolism, endocarditis and bleeding. Clinicians and patients often mistakenly believe they no longer require secondary prophylaxis following cardiac surgery. They are still at risk of recurrences of ARF and secondary prophylaxis must often continue for life.

## **Long-term postoperative management**

Heart valve surgery requires regular long-term follow-up. Ideally, this should be done in a centre equipped with echocardiography. Individuals who have had conservative valve procedures, such as valvotomy or valve repair, require close observation to detect re-stenosis or a recurrence of valve regurgitation, and to ensure secondary prophylaxis is administered regularly. It is also important to monitor LV and prosthetic function. If echocardiography is not available, patients should be referred back to the surgical centre or local medical specialist if they develop any of the following:

- recurrent symptoms
- evidence of heart failure
- a new regurgitant murmur
- any thromboembolic episode
- signs and symptoms suggesting endocarditis.

## **Anticoagulation needs to be monitored following replacement with mechanical valves.**

Good anticoagulation management requires standardized anticoagulation measurement, using the International Normalised Ratio (INR). The dose of anticoagulation required depends on the individual and the type of prosthetic heart valve used. Regular monitoring of the INR and maintaining it within the therapeutic range may be difficult for people living in areas where health services are difficult to access.

All people requiring anticoagulation must receive regular care from a medical specialist.[47]

**All patients who receive a prosthetic valve replacement must remain on BPG as secondary prophylaxis for life.**

## **Notification**

- ARF is currently notifiable to MoHMS. RHD is not currently notifiable in Fiji.
- Notification of ARF in Fiji is to the RFIS and to Health Information System
- The Divisional Coordinator will liaise with clinic/hospital nurses/SMOs to ensure reporting is undertaken for all new and suspected patients.

## **Echocardiography screening**

### **Screening for Rheumatic Heart Disease**

The WHO now recommends active case finding for RHD as a tool for estimating the disease burden, and also for identifying cases in areas with a high prevalence of RHD.[51]WHO Global Programme on RHD undertook auscultatory screening of over one million children. In some regions, this was augmented by echocardiography to confirm the diagnosis of RHD. The sensitivity of cardiac auscultation is highly dependent on the skill of the operator, and the specificity of auscultation for rheumatic carditis is low. From 2007 there has been considerable international endeavour studying the use of echocardiographic screening for RHD.[34, 38, 52-57]It has been established that:

- Echocardiography is more accurate than auscultation for screening. [11, 38, 53]
- By using portable echocardiography, it is feasible to screen for RHD in school based programmes.
- Echocardiography is acceptable to populations at risk. [58]
- Previous WHO and WHO-NIH criteria resulted in over-diagnosis of RHD.[55]

An international group of researchers (led from New Zealand and Australia) achieved standardisation of criteria for the minimal diagnosis of RHD (endorsed and published as the WHF guidelines 2012) using the best available evidence (echocardiographic, surgical and pathological descriptions of what does or does not constitute RHD).[44]

However the natural history of echocardiographically detected RHD is still being clarified. The Australian follow up study has shown that even borderline RHD carries an 8-9 times greater chance of RHD progression over a 3.5 year period compared to a control group.[59] Milder valve lesions, which are often asymptomatic and thus the most common lesions that have been detected with echo screening, have the most to gain from secondary prophylaxis. [60-62]

Echocardiographic screening is labour intensive and requires a skilled group of echocardiographers to perform the heart scans, interpret the echocardiograms and clinicians to counsel those with positive tests. Good secondary prophylaxis needs to be in place as those with unequivocal RHD are started on penicillin. The utility of echocardiographic detection as a contributor to RHD control in Fiji is currently being evaluated and a recommendations paper will be developed in 2017.

The Fiji National Strategic plan 2016-20 reportable indicator for RHD is the percentage of primary aged school students screened for RHD states under Priority area 2: 4.2.42014 baseline (21%) with a 2020 goal  $\geq 47\%$  (the screening method is not identified) The MoHMS Fiji Child and Adolescent Health Strategy 2016-2020 lists additional national indicators for ARF and RHD (MoHMS draft version 2016). When the screening recommendations are agreed upon both the Annual Corporate Plan 2016 and Fiji RHD Policy (2015) may require amendment.



## Appendices

### Appendix 1: WHF criteria for the minimal echocardiographic criteria for the diagnosis of RHD [44]

#### 1. Echocardiographic criteria for individuals $\leq 20$ years

##### Definite RHD (Either A, B, C or D)

- A. Pathological MR and at least 2 morphological features of RHD of the mitral valve
- B. MS mean gradient 4mmHg (*NB: Congenital mitral valve abnormalities must be excluded*)
- C. Pathological AR and at least 2 morphological features of RHD of the aortic valve (*NB: Bicuspid aortic valve and dilated aortic root must be excluded*)
- D. Borderline disease of both Aortic and Mitral Valves\*

##### Borderline RHD (Either A, B or C)

- A. At least 2 morphological features of RHD of the mitral valve without pathological MR or MS
- B. Pathological MR
- C. Pathological AR

##### Normal echocardiographic finding of (all of A, B, C and D)

- A. MR that does not meet all four Doppler criteria (physiological MR)
- B. AR that does not meet all four Doppler criteria (physiological AR)
- C. An isolated morphological feature of RHD of the mitral valve e.g. Valvular thickening, without any associated pathological stenosis or regurgitation
- D. Morphological feature of RHD of the aortic valve e.g. Valve thickening, without any associated pathological stenosis or regurgitation

#### 2. Echocardiographic criteria for individuals $\geq 20$ years

- A. Pathological MR and at least 2 morphological features of RHD of the mitral valve
- B. MS mean gradient 4mmHg (*NB: Congenital mitral valve abnormalities must be excluded*)
- C. Pathological AR and at least 2 morphological features of RHD of the aortic valve in individuals  $< 35$  years of age only (*NB: Hypertension, bicuspid aortic valve and dilated aortic root must be excluded*)
- D. Pathological AR and at last 2 morphological features of RHD of the mitral valve

\*Combined AR and MR in high prevalence regions and in the absence of CHD is regarded as rheumatic AR, aortic regurgitation; AV, aortic valve; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve.

## Appendix 2 : ARF and RHD Programme Key Performance Indicators

1. Epidemiology and reporting		
1.1	Yearly age specific incidence rates of all episodes, and of first episodes of ARF according to gender and ethnicity	0-4, 5-14, 15-24, 35-44 and >44 years
	1.1.1 Gender	Male, Female, Not-stated/ Inadequately described
	1.1.2 Ethnicity	Indigenous Fijian (iTaukei), Indo-Fijian, Other
1.2	Proportion of all recorded ARF episodes classified as recurrences	
1.3	Rates of ARF recurrences per 100 patient-years	
1.4	Number of deaths and age-standardised rates of mortality due to ARF and RHD in the previous calendar year by ethnicity (refer 1.1.2)	
	1.4.1 Number of deaths during pregnancy of ARF and RHD patients and rate of maternal mortality due to ARF and RHD	
1.5	Yearly age- specific (refer to 1.1) and overall incidence of RHD by ethnicity (refer to 1.1.2) and segregated by location, and method found.	<ul style="list-style-type: none"><li>- all recorded RHD cases</li><li>- cases classified as mild</li><li>- cases classified as moderate</li><li>- cases classified as severe</li></ul>
1.6	Yearly age- specific (refer to 1.1) prevalence by ethnicity (refer to 1.1.2)	<ul style="list-style-type: none"><li>- all recorded RHD cases</li><li>- cases classified as mild</li><li>- cases classified as moderate</li><li>- cases classified as severe</li></ul>
1.7	Proportion of newly registered cases of ARF and RHD with an initial recorded diagnosis being established RHD (rather than ARF)	
2. Secondary Prophylaxis		
2.1	Proportion of all people indicated for secondary prophylaxis who are registered to receive BPG by age group (refer 1.1)	
2.2	Median percentage of all scheduled BPG actually delivered	
2.3	Proportion of people indicated for BPG secondary prophylaxis who received > 80 %, and 100 % of scheduled doses in the previous calendar year	
3. Quality management		
3.1	Proportion of all registered ARF and RHD cases classified as mild, moderate or severe and inactive	
3.2	Proportion of ARF cases with probable ARF diagnosis	



## Appendix 3 : ARF/RHD Patient Registration Form (version 25/11/2015) ARF/RHD Patient Registration Form

*For Registration of Newly Diagnosed and Suspected Cases of ARF and RHD and ARF Recurrences. Note for nurses: copy the patient information from the patient book or injection book onto the relevant sections of this form.*

<b>REFERRAL DETAILS</b>	
Date Referred: ...../...../..... Referred By: ..... Role: .....	
Referral source: <input type="checkbox"/> GP <input type="checkbox"/> Hospital <input type="checkbox"/> Health Centre <input type="checkbox"/> Nursing Station <input type="checkbox"/> Other.....	
Name of referral facility.....	
<b>PATIENT DETAILS:</b> <span style="float: right;">NHN: .....</span>	
First name: ..... Last name: ..... Date of Birth: ...../...../.....	
Sex: <input type="checkbox"/> Male <input type="checkbox"/> Female Ethnicity: <input type="checkbox"/> iTaukei <input type="checkbox"/> Fijians of Indian Descent <input type="checkbox"/> Other (specify) .....	
Division: <input type="checkbox"/> Central <input type="checkbox"/> Eastern <input type="checkbox"/> Western <input type="checkbox"/> Northern	
Contact person 1: ..... Relationship: ..... Phone(s): .....	
Contact person 2: ..... Relationship: ..... Phone(s): .....	
Address at diagnosis: ..... Current address (if different): .....	
Current school (if attending): .....	
Primary Treatment centre: ..... Secondary Treatment Centre .....	
<b>CLINICAL COURSE AND OUTCOME</b>	
Diagnosis date: ...../...../..... Diagnosed by: <input type="checkbox"/> GP <input type="checkbox"/> MO <input type="checkbox"/> NP Name: .....	
Diagnosis Made in Pregnancy or Post-delivery: <input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Unknown	
Benzathine: <input type="checkbox"/> 3 weekly <input type="checkbox"/> 4 weekly Start date: ...../...../..... <input type="checkbox"/> Other prophylaxis: .....	
Other Medications: Warfarin: <input type="checkbox"/> Yes <input type="checkbox"/> No Heart Failure medications: <input type="checkbox"/> Yes <input type="checkbox"/> No	
Admitted to Hospital: <input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Unknown	
Date of Hospitalisation ...../...../..... Hospital: .....	
Died: <input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Unknown Date of Death ...../...../..... Cause of Death: .....	
<b>ARF DIAGNOSIS SUSPECTED?</b> <input type="checkbox"/> YES: First Diagnosis <u>OR</u> <input type="checkbox"/> YES: Recurrence <input type="checkbox"/> No	
Strep Evidence: <input type="checkbox"/> High ASOT for age <input type="checkbox"/> GAS throat swab	
Major Criteria: <input type="checkbox"/> Carditis on ECHO (also record as RHD below) <input type="checkbox"/> Clinical carditis <input type="checkbox"/> Chorea	
<input type="checkbox"/> Monoarthritis or polyarthritis or polyarthralgia <input type="checkbox"/> Erythema Marginatum <input type="checkbox"/> Subcutaneous nodules	
Minor Criteria: <input type="checkbox"/> Monoarthralgia <input type="checkbox"/> Fever $\geq 38^{\circ}\text{C}$ <input type="checkbox"/> Prolonged PR <input type="checkbox"/> ESR $\geq 30$	
<b>RHD DIAGNOSIS SUSPECTED?</b> <input type="checkbox"/> Yes <input type="checkbox"/> No <b>RHD PREVIOUSLY DIAGNOSED?</b> <input type="checkbox"/> Yes	
Evidence: <input type="checkbox"/> Murmur <input type="checkbox"/> Heart failure <input type="checkbox"/> CXR <input type="checkbox"/> ECG <input type="checkbox"/> Other.....	
ECHO Result: <input type="checkbox"/> No RHD <input type="checkbox"/> Borderline RHD <input type="checkbox"/> Mild RHD <input type="checkbox"/> Moderate RHD <input type="checkbox"/> Severe RHD <input type="checkbox"/> Awaiting report <input type="checkbox"/> Not done	
<b>FOLLOW-UP ARRANGED – Referrals made</b>	
<input type="checkbox"/> Specialist/Paediatrician Date: ...../...../..... Where: .....	
<input type="checkbox"/> Dentist Date: ...../...../..... Where: .....	
<input type="checkbox"/> Echo review Date: ...../...../..... Where: .....	
<input type="checkbox"/> Other Date: ...../...../..... Specify: .....	

Date Form completed Date: ...../...../.....

## Appendix 4 : Standard Operating Procedure for ARF/RHD Patient Registration Form Completion

When is form to be completed	New ARF or RHD patient who attends clinic or health facility (Note; does not include patients visiting from other clinics for injection, change of RHD diagnosis or address/contact details) Suspected case of ARF attends clinic – may be known or unknown patient
Who completes form	Clinic/treatment room staff/MO's and NP's
How is the form collected	Designated staff e.g. SOPD nurse or RHD Liaison submits form to Divisional RHD Coordinator (see bottom of form for details) – forms can be sent together with Injection form or in between quarterly injection data reporting or fax directly to RHD programme
Referral Details	Please complete this section with referring doctor or nurse details, including name, location and type of facility and date patient was referred
Patient Details	Patient identification details must be completed for registration and patient follow-up. Please complete all fields. Primary Treatment Center refers to health facility where patient receives most injections. Secondary clinic refers to other health facility that the patient attends to receive treatment.
Clinical Course and Outcome	<ul style="list-style-type: none"> <li>• Diagnosis date is the date that the patient received diagnosis from a MO, GP or NP.</li> <li>• If diagnosis was made in pregnancy this section must be completed.</li> <li>• Benzathine schedule must be ticked either 3 weekly or 4 weekly- to aid calculation of patient adherence.</li> <li>• Please complete the date the patient was first prescribed BPG if known.</li> <li>• Other medications: complete if patient has a warfarin order. Other heart failure medications refer to diuretics, beta- blockers, etc.</li> <li>• Hospitalization details should be completed if the patient has been hospitalised at time of diagnosis.</li> <li>• If the patient has died please complete date of death and record cause of death as it appears on the death certificate.</li> </ul>
ARF diagnosis suspected	Please check each box in this section that is applicable – if not applicable or not available then leave box unchecked
RHD diagnosis suspected	Please check each box in this section that is applicable – if not applicable or not available then leave box unchecked. Tick "RHD PREVIOUSLY DIAGNOSED" box ONLY if patient is a known case of RHD with a suspected or known case of ARF
Follow-up arranged	Please complete this section with booking that have been scheduled, including date and location for review
Sending the form to RHD team	<ul style="list-style-type: none"> <li>• For the East and Central please send the completed ARF/RHD patient registration form to the Central/Eastern RHD Divisional Coordinator.</li> <li>• For the North please send the completed ARF/ RHD patient registration form to the Northern RHD Divisional coordinator</li> <li>• For the West please send the completed form to the Western RHD Divisional coordinator- contact details are on the registration form.</li> </ul>

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