National Protocol
For Diagnosis, Management and Prevention of Acute Rheumatic Fever and Rheumatic Heart Disease (National RHD Protocol)

(Protocal for Health Care Professionals Working in Basic Health Services Centers, Basic Hospitals and District Hospitals)

Government of Nepal
Ministry of Health and Population
Department of Health Services
Epidemiology and Disease Control Division
Teku, Kathmandu, Nepal

Diagnosis, Management and Prevention of
Rheumatic Fever and Rheumatic Heart Disease
(National RHD Protocol)
(Protocol For Health care professionals working in Basic Health Services Centers, Basic Hospitals and District Hospitals)

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ABBREVIATIONS

ACEI:	Angiotensin Converting Enzyme Inhibitor
AF:	Atrial Fibrillation
AHA:	American Heart Association
AR:	Aortic Regurgitation
ARB:	Angiotensin Receptor Blocker
ARF:	Acute Rheumatic Fever
AS:	Aortic Stenosis
ASOT:	Antistreptolysin O Titre
AV:	Atrioventricula
BHSC:	Basic health service center
BH:	Basic hospital
BID:	Two times daily
BMV:	Balloon Mitral Valvotomy
BP:	Blood Pressure
BPG:	Benzathine Penicillin G
BTP:	Bacterial tonsillopharyngitis
CCB:	Calcium Channel Blocker
CCF:	Congestive Cardiac Failure
CRP:	C-reactive Protein
CXR:	Chest X-ray
DNAaseB:	Deoxyribonuclease B
DH:	District Hospital
ECG:	Electrocardiogram
Echo:	Echocardiogram
ENT:	Ear, nose and throat
ESR:	Erythrocyte Sedimentation rate
GAßHS:	Group A ß-hemolytic Streptococci
GAS:	Group A Streptococcus
INR:	International Normalized Ratio
IU:	International Unit

IM:	Intramuscular
IV:	Intravenous
JVP:	Jugular Venous Pressure
LA:	Left Atrium
LMWH:	Low Molecular Weight Heparin
LV:	Left Ventricle
MR:	Mitral Regurgitation
MS:	Mitral Stenosis
NCD:	Non- Communicable Disease
NHF:	Nepal Heart Foundation
NOAC:	New Oral Anticoagulant
OD:	Once Daily
PASP:	Pulmonary Artery Systolic Pressure
PCM:	Paracetamol
PO:	By mouth
PS:	Pulmonary Stenosis
PST:	Penicillin Sensitivity Tes
PTAV:	Percutaneous Transluminal Aortic Valvuloplasty
RA:	Right Atrium
PTMC:	Percutaneous Transvenous Mitral Commissurotomy
RHD:	Rheumatic Heart Disease
QID:	Every 6 hours (4 times per day)
RF:	Rheumatic Fever
RV:	Right Ventricle
SGNHC:	Shahid Ganga Lal National Heart Centre
SOB:	Shortness of Breath
TDS:	Three times daily
UFH:	Unfractionated Heparin
WBC:	White Blood Cell
WHF:	World Heart Federation
WHO:	World Health Organization

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CHAPTER - 1

OVERVIEW

Background

Rheumatic heart disease (RHD) is a non-communicable disease, although the beginning of RHD is infectious in origin. There are three stages in the development of RHD. It begins with acute tonsillopharyngitis caused by Group A Beta Hemolytic Streptococci (GAS) (**first stage**), extends to acute rheumatic fever (ARF) (**second stage**), then ends with heart valve damage known as RHD (**third stage**).

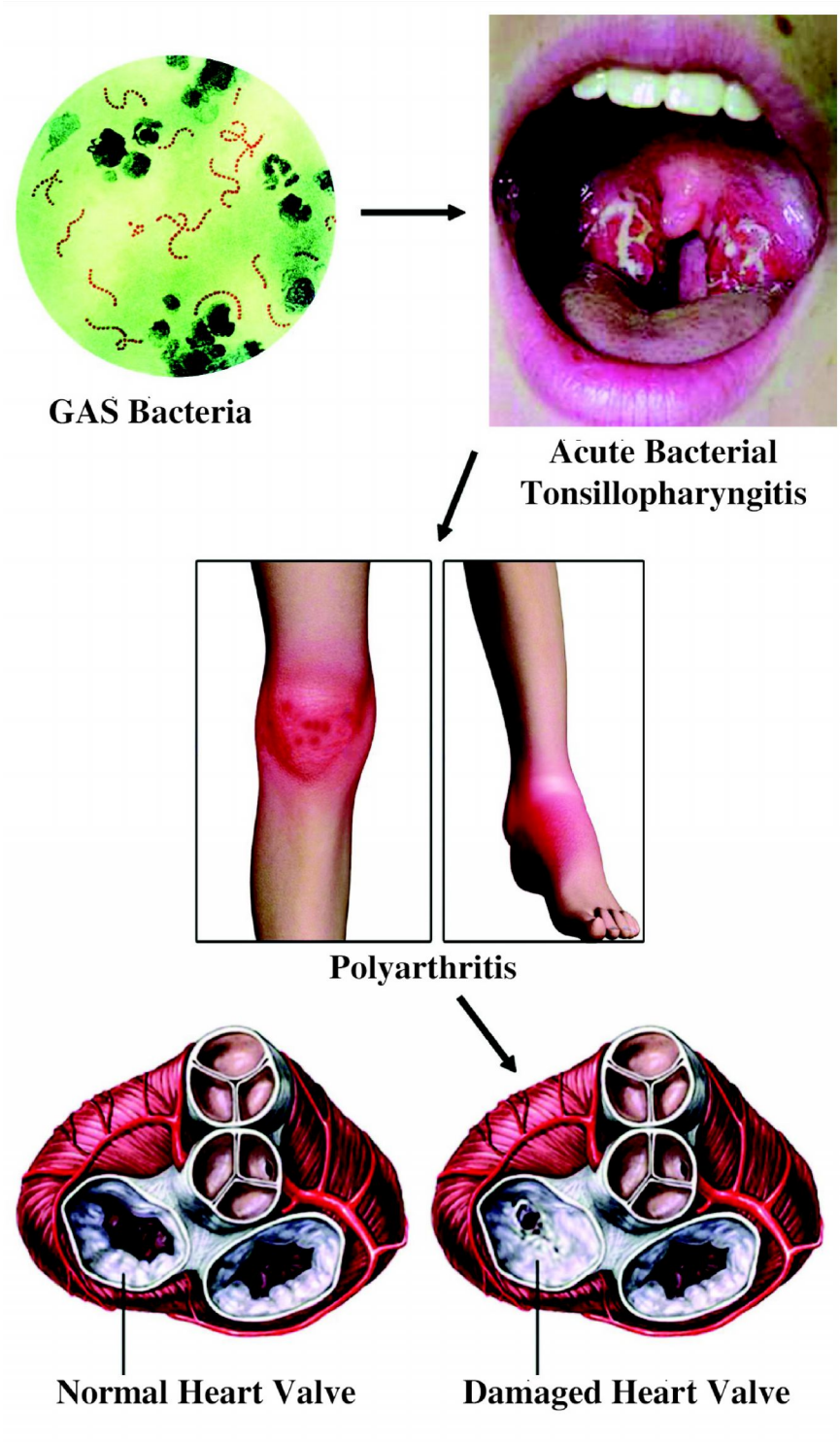


Figure 1: Development pathway of RHD

Bacterial tonsillopharyngitis (BTP) is a common problem in children and young adults and a gateway for development of ARF and RHD. About one third of all bacterial tonsillopharyngitis are caused by GAS. ARF is a systemic inflammatory disorder which is developed as a result of autoimmune response to untreated and repeated episodes of GAS tonsillopharyngitis. ARF ultimately leads to

RHD which is a long-term condition with heart valve damage. Thus, RHD is directly linked with bacterial tonsillopharyngitis. Public knowledge about this link is very low.

RF most often occurs between 10 to 21 days after a GAS infection and most commonly affects the heart (carditis), large joints (arthritis or arthralgia), brain (chorea), skin (erythema marginatum). and subcutaneous tissues (subcutaneous nodules). The first episode of RF is usually seen in children aged 5 to 14 years; recurrent episodes are most common within 1 year of the first episode but can occur throughout the life course. RHD most commonly starts in childhood with a diagnostic peak in young adults aged 20 to 39 years. RHD can lead to death or lifelong disability, however, effective early intervention can prevent premature morbidity and mortality. (1)

RHD affected an estimated 55 million people globally and caused 360,000 deaths in 2021 (2). In the twentieth century, the incidence of ARF and the prevalence of RHD declined substantially in Europe and North America, and in other high-income settings. However, the gains have not been equitably distributed globally and many regions including sub-Saharan Africa, the Middle East, Central and South Asia, tropical Latin America and the South Pacific continue to have endemic ARF and RHD. The prevalence of RHD is estimated to peak between the ages of 20 and 29 years, declines steadily until around 50 years when it then remains relatively stable (3). There is a higher prevalence of RHD among women across nearly all world regions (3).
Countries in Southeast Asia, including Nepal, are endemic with RHD. However, RHD related mortality rates have shown improvement in several countries over the past two decades. (4) According to the GBD data, RHD mortality in Pakistan and India declined between 2011 and 2021, from 12.28 to 10.28, and 13.97 to 11.74 deaths per 100,000 population, respectively. (4) In contrast, Nepal's RHD mortality rate has remained relatively unchanged, fluctuating slightly between 12 and 12.1 deaths per 100,000. (4)

RHD is a significant public health problem in Nepal taking away lives of thousands of children and young adults annually. RHD Prevalence in Nepal has been estimated mainly from surveys of school-going children using heart screening by auscultation method and it varies from 0.9 to 7.32 per thousand school children. (5-10). The prevalence of RHD varied across different districts, provinces and ecological zones of Nepal. A study conducted in 25 districts of Nepal (11) covering all 7 provinces and the three ecological zones showed the highest prevalence in Karnali province (3.45/1000) and the lowest in Bagmati province (1.38/1000). The prevalence was highest in the Terai zone (2.89 per 1000) and lowest in the northern mountain zone (1.56 per 1000).
Echocardiographic screening showed ten times more cases of RHD compared with auscultation. (12) Although few regional studies have shown the burden of RHD to be high in Nepal, nationwide prevalence study is lacking. Estimated prevalence indicates Nepal to be in a high endemic RHD zone. Echocardiographic evidence of subclinical RHD has been documented as 1 in 100 children in Nepal and represents a serious public health challenge. It is more common in girls, older age group children and under-privileged children. (12)
RHD is diagnosed by clinical examination and imaging technique like echocardiography. RHD is clinically diagnosed based on the detection of a pathological heart murmur during auscultation. Echocardiography is used to determine the source and cause of murmurs discovered during auscultation. Comprehensive echocardiography screening is the best tool for population screening due to its higher

sensitivity and its ability to diagnose subclinical diseases.

In Nepal a gross estimate made by Nepal Heart Foundation (NHF) revealed:

- More than one million existing RHD cases
- Around 30,000 people get affected and 3000 people die prematurely from RHD every year
- 10 % of all medical admissions are for RHD
- 37% of all cardiac surgeries at the tertiary heart centers are related to RHD
- 10% of all trans-catheter interventions in tertiary heart care centers are related to RHD
- More than 1000 patients go for heart valve surgery annually
- More than 20% maternal death during delivery of child is related to RHD

According to the World Heart Federation (WHF) criteria**,** the population of Nepal falls under the category of moderate to high-risk for RF/RHD (ARF incidence of $\geq$2per 100,000 school-age children per year or RHD prevalence of $\geq$1 per 1000 population). (13)

Introduction

This protocol on RHD has been prepared to enhance effective, efficient, yet feasible RHD care and prevention efforts in Nepal. This protocol is intended for use by a wide group of health professionals working in Basic Health Service centers (BHSC), Basic hospitals (BH) and district hospitals (DH).

CHAPTER - 2

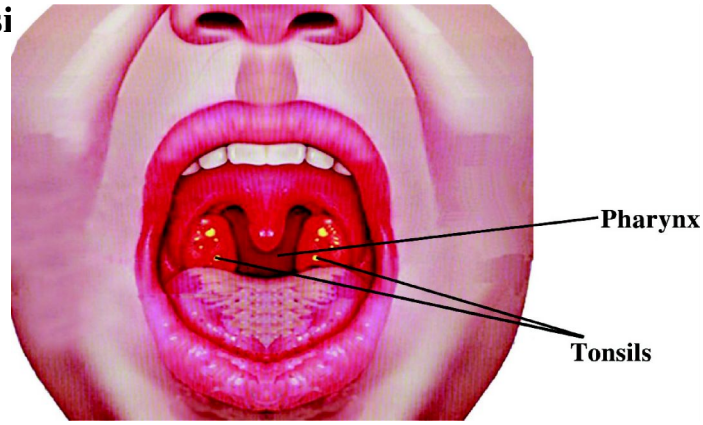
DIAGNOSIS, MANAGEMENT AND PREVENTION OF BACTERIAL TONSILLOPHARYNGITIS

Bacterial Tonsillopharyngitis

Introduction

- Throat infection (tonsillopharyngitis) is mostly caused by virus and bacteria
- The most common bacteria causing sore throat is Group A beta hemolytic streptococcus (GAS)
- Children of age 5 to 15 years are at greater risk of being infected by GAS
- Untreated and repeated GAS tonsillopharyngitis may lead to development of ARF and RHD

Symptoms and signs of bacterial tonsi

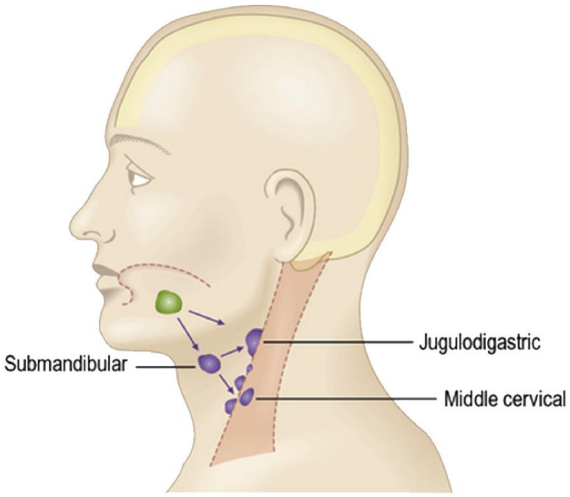


Symptoms:

1. High Fever
2. Pain in throat
3. Difficulty in swallowing
4. Bad breath

Signs:

1. Enlarged red tonsils
2. Redness of pharynx
3. Pus like Exudates on pharynx and tonsils
4. Bleeding spots
5. Enlarged lymph nodes of the neck (commonly the Jugulodigastric L/N are enlarged which are found just below the angle of mandible, deep to the sternocleidomastoid muscle)



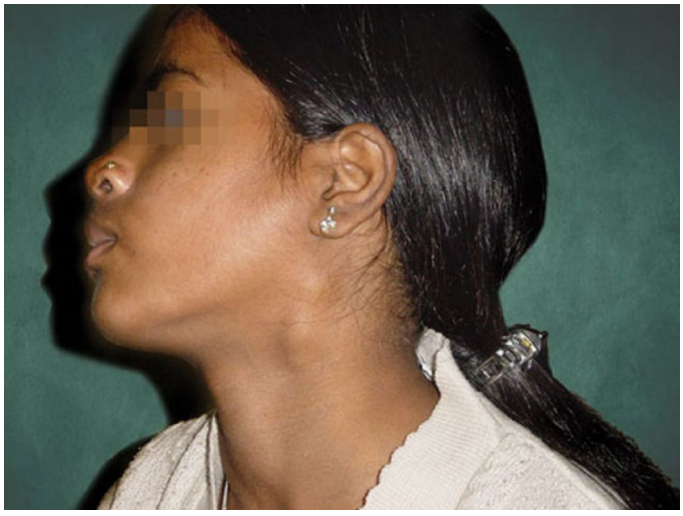


Figure 3: Location of Jugulodigastric lymph nodes

How to differentiate Bacterial and Viral Tonsillopharyngitis?

Sign and Symptoms	Bacterial	Viral
Throat Pain	**Present +++**	**Present** +
Difficulty Swallowing	**Present +++**	Absent or may be present
Enlarged and Red tonsils	**Present +++**	**Present +**
White or yellow spots on tonsils and/or pharynx	**Present**	Absent
Enlarged lymph nodes of neck	**Present**	Absent
Bleeding spots	**Present+++**	Absent or may be present +
Cough	Absent	**Present**
Hoarseness	Rare	**Common**
Sneezing	Absent	**Present**
Runny Nose	Absent	**Present**
Fever	**Moderate-High grade (>100.4 F)**	**Low-grade (99 - 100.4F) or Absent**

How to confirm the diagnosis of bacterial tonsillopharyngitis?

The diagnosis can be confirmed on the following basis:

- Positive Throat swab culture OR
- Positive Rapid Strep Test

NB: It may not be feasible to do these tests in the Basic Health Service settings. It is not advised to wait for the confirmation of the diagnosis and delay initiation of treatment.

How to treat bacterial tonsillopharyngitis? (Table 1)

Table 1: Treatment of bacterial tonsillopharyngitis

Drugs	Route	Dose	Frequency	Duration
Benzathine Penicillin G	Deep IM	600,000 units (wt. ≤27Kg) 1,200,000 units (wt. >27 Kg)	Single dose	-
Penicillin V (Phenoxymethyl Penicillin)	Oral	250mg (wt ≤27kg) 500mg (wt > 27kg)	TDS (8 hourly)	10 days
Amoxycillin	Oral	Children (10-15mg/ kg/dose) Adults 500mg	TDS (8 hourly)	10 days
Azithromycin	Oral	10 - 12mg/kg/day Max dose 500mg	OD (once daily)	5 days

NB: Penicillin (Injection or Oral) should be the first line drug

Recommendations for Management of Tonsillopharyngitis

1. Diagnose bacterial tonsillopharyngitis on the basis of clinical signs and symptoms.
2. Suspected bacterial tonsillopharyngitis - treat with antibiotic. Penicillin should be the drug of choice, Amoxicillin and Azithromycin may be used as alternatives.
3. Doubtful or borderline or if you cannot decide clinically whether it is bacterial or viral - treat with antibiotic.
4. Mixed type - bacterial and viral (cases having signs and symptoms of both bacterial and viral infection) - Treat with antibiotic.
5. Suspected Viral - don't treat with antibiotic. Treat with Paracetamol and saline gargling.

Prevention of bacterial tonsillopharyngitis in children
Bacteria can infect and spread from one child to another by:

- Coughing and sneezing
- Not washing hands
- Sharing foods and drinks
- Poor nutrition
- Poor personal hygiene
- Overcrowding

The following precautions should be taken to prevent BTP in children:

- Cover mouth while coughing and sneezing
- Wash hands frequently
- Avoid sharing consumed foods and drinks
- Good nutrition to build up immunity
- Brush teeth twice daily
- Isolation of infected case

Action protocol on diagnosis & management of bacterial tonsillopharyngitis
Action 1: Ask
Ask about the symptoms of throat infection. The following symptoms are common for BTP:

- throat pain,
- difficulty in swallowing
- fever
- Absence of cough

Action 2: Assess (Examine the throat and neck)
Look for signs of Bacterial infection

- Enlarged and red tonsils,
- Redness of pharynx,

- Pus like exudates on pharynx and tonsils,
- Bleeding spots
- Enlarged lymph nodes of the neck (jugulodigastric lymph nodes)
- Fever (Check temp)

Action 3: Estimate

- Apply clinical decision rule to differentiate bacterial from viral infection
- Bacterial- throat pain, difficulty in swallowing, enlarged cervical lymph nodes, high grade fever
- Viral- runny nose, sneezing, cough, hoarseness, low grade fever
- Most of the time no laboratory test is needed to establish the diagnosis

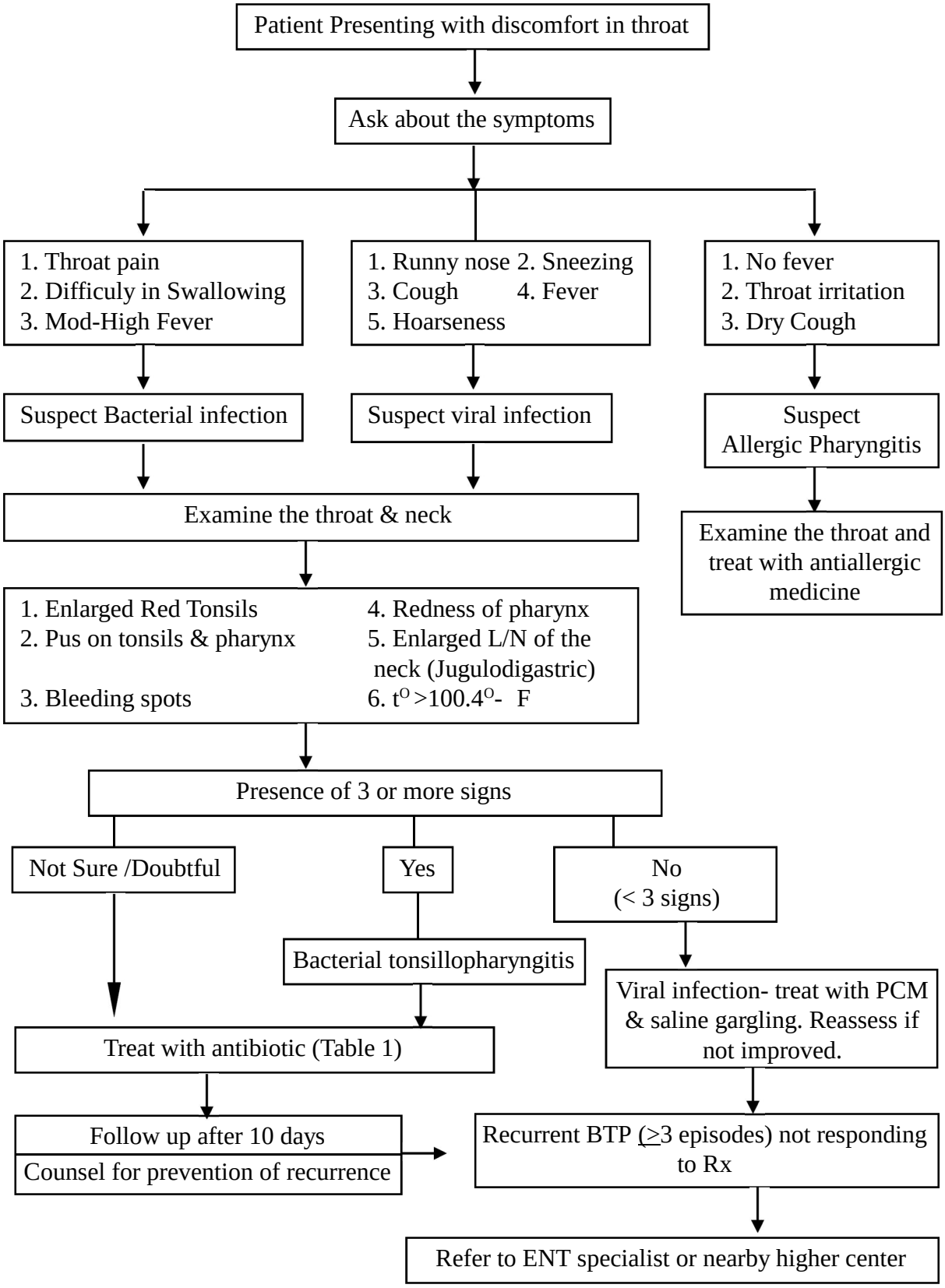
Action 4: Treat as necessary and Counsel

Treat as per bacterial tonsillopharyngitis treatment guidelines (**Table1**)

Action 5: Refer if necessary

Recurrent throat infection ($\geq$ 3 episodes) not responding to antibiotics - refer to ENT specialist (if available) or higher center

Algorithm for Suspected Bacterial Tonsillopharyngitis (BTP)



NB: Untreated bacterial tonsillopharyngitis may lead to development of ARF.
5-15 Years age group has high risk for development of ARF

CHAPTER - 3

DIAGNOSIS, MANAGEMENT AND PREVENTION OF ACUTE RHEUMATIC FEVER (ARF)

Acute Rheumatic Fever

Introduction:

- Acute rheumatic fever (ARF) is a delayed autoimmune response to an untreated Group A â Hemolytic streptococcus (GAS) infection, resulting in inflammation of specific connective tissues.
- ARF develops about 2-3 weeks after the onset of a GAS tonsillopharyngitis.
- 0.3 to 3% of GAS sore throat progress to ARF if left untreated.
- Approximately half of the ARF cases progress to RHD.
- ARF may involve the heart, joints, central nervous system, skin and subcutaneous tissue.
- ARF occurs commonly between the ages of 5-15 years.
- The illness usually lasts up to 3 months and resolves without treatment if heart is not affected.
- With treatment the symptoms resolve within 1-2 weeks.
- ARF can occur repeatedly in people who continue to be exposed to group A â Hemolytic streptococci in their environment.
- 50% cases of ARF do not give history of bacterial tonsillopharyngitis.

Risk Factors of ARF

- Overcrowding
- Limited access to health care
- Heredity
- Poor standard of housing
- Poor nutrition
- Age 5-15 years

Clinical manifestations of ARF

Major manifestations (more specific)

1. **Carditis** (clinical and/or subclinical i.e. echo diagnosed): presents with symptoms of heart failure such as shortness of breath (SOB), orthopnea, paroxysmal nocturnal dyspnea (PND), palpitation etc
2. **Arthritis** (migratory polyarthritis or Monoarthritis): single or multiple large joints (shoulder, elbow, wrist, hip, knee, ankle etc) involvement causing swelling, pain, redness and hotness of affected joints, migrating in nature, non-suppurative, self - limiting, without causing residual deformity. OR **Polyarthralgia** (after excluding other causes): multiple joints pain without swelling.
3. **Sydenham's Chorea:** presents with Jerky, purposeless, involuntary movements of hands, legs and other parts of the body. The movements disappear when asleep. Sometime hemichorea is observed with movements limited to one side of the body. Usually this is a late presentation seen when other signs and symptoms have disappeared.
4. **Erythema marginatum** (skin rash): presents with macular rashes over the trunk and proximal parts of limbs, which are circular, painless, non-itching, white area with pink margins. These rashes do not appear on face, palms and soles.
5. **Subcutaneous nodules:** small, painless, mobile pea sized nodules seen under the skin, over the extensor surface of joints, bony prominences or tendons

Minor manifestations (less specific)

1. **Fever** (≥38^0 C or ≥100.4 ^oF)
2. **Monoarthralgia:** single joint pain. Large joints of hands or legs are commonly involved
3. **Prolonged P-R** interval in ECG (> 0.20 seconds)
4. **Raised ESR** (≥ 30 mm/h) OR Raised **CRP** (≥3.0mg/dl)

Supporting evidence of preceding GAS Infection. (Essential criteria)

- Raised Anti-Streptolysin O titre (ASOT >200 units)
- Raised AntiDNase B titre Positive
- Positive throat swab culture for GAS positive

(NB: ASOT test is commonly available and more feasible in our health facilities)

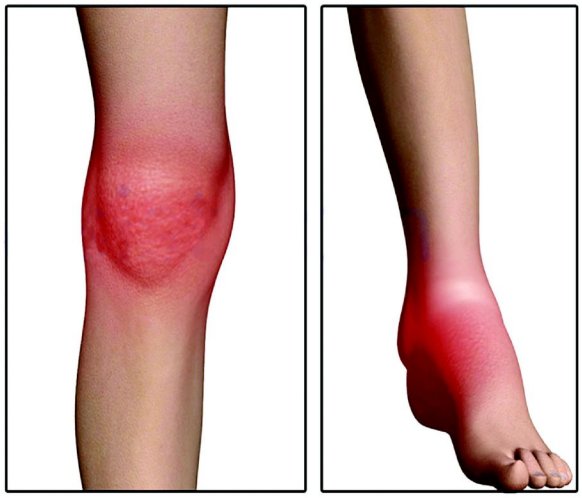


Figure 4: Arthritis

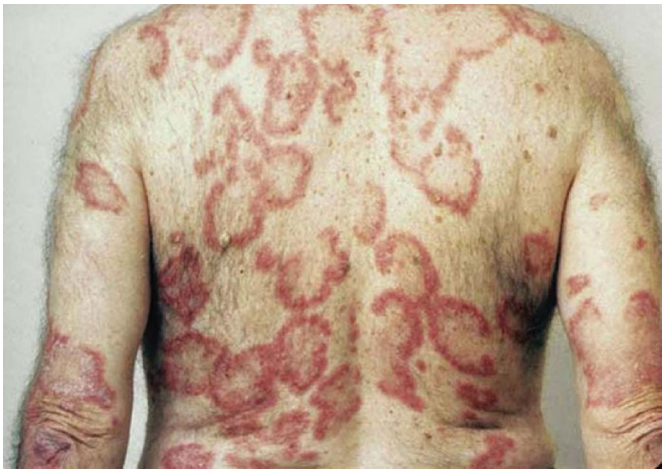


Figure 5: Erythema Marginatum

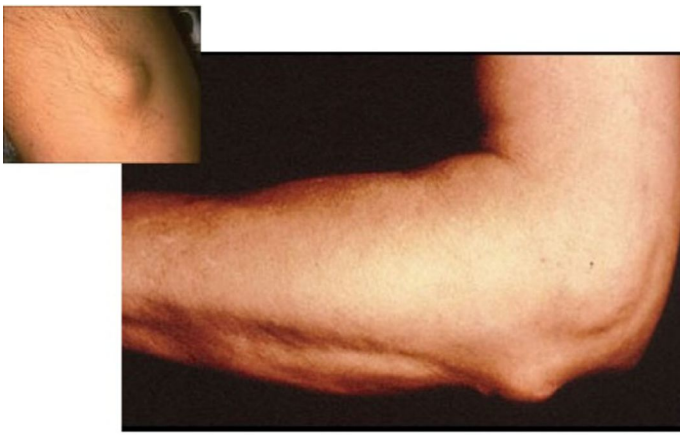


Figure 6: Subcutaneous Nodule

Figure 7: Chorea

How to Diagnose ARF?

There is no specific diagnostic test for acute rheumatic fever.

Diagnosis of ARF is made using the **Jones criteria (2015 modification by AHA) for high-risk populations (14) (Table 2)**

Jones criteria include Major manifestations, minor manifestations and supporting evidence of preceding GAS infection as described above in clinical manifestations of ARF.
The diagnosis of ARF can be confirmed in the presence of following situations:

Tables 2: Diagnosis of rheumatic fever based on revised Jones criteria updated in 2015.

	Mod - High risk populations (incidence of ARF ≥2/100,000 school-aged children per year or prevalence of RHD≥1/1,000 population)
A. Major clinical manifestations	**Carditis:** Clinical and/or subclinical (echocardiographic valvulitis)
	Arthritis: Monoarthritis OR polyarthritis OR polyarthralgia
	Chorea
	Erythema marginatum
	Subcutaneous nodules
B. Minor Clinical manifestations	Monoarthralgia
	Fever (≥380C)
	Raised ESR (≥30 mm in the first hour) and /or CRP (≥3 mg/dL)
	Prolonged PR interval (> 0.20 seconds)
C. Supportive evidence of Preceding GAS infection	Positive throat culture or rapid streptococcal antigen test or elevated streptococcal antibody titer (ASOT).
Diagnosis of initial attack of ARF	2 major manifestations OR 1 major plus 2 minor manifestations Plus supportive evidence of preceding GAS infection
Diagnosis of recurrent attack of	2 major manifestations OR 1 major plus 2 minor manifestations OR 3 minor manifestations plus supportive evidence of preceding GAS infection

NB: In case of Chorea, presence of Chorea alone is sufficient for the diagnosis of ARF
(ARF: acute rheumatic fever: CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; GAS: group A Streptococcus; RHD: rheumatic heart disease)

Any of the following is Acute Rheumatic fever

Presence of 2 major manifestations plus any evidence of preceding GAS infection

OR

Presence of 1 major manifestation, 2 minor manifestations plus any evidence of preceding GAS infection

OR

Presence of Rheumatic (Sydenham's) Chorea

OR

Presence of 2 major or 1 major and 2 minor or 3 minor manifestations plus any evidence of preceding GAS infection (In case of Recurrent ARF)

Clinical conditions of ARF

a. Recurrent ARF: A repeated episode of acute rheumatic fever following another GAS infection. (Diagnosis is made by using Jones Criteria 2015)

b. Subclinical Carditis: When the clinical examination is normal but the echocardiogram shows valve damage. (Diagnosis is made by echocardiography screening)

c. Indolent Carditis: Carditis of slow progression or insidious onset. The patient is mostly without symptoms but may have heart murmur on auscultation and usually presents months after the onset of ARF. (Diagnosis is made by echocardiography screening)

Classification of ARF

Definite ARF: acute presentation which fulfils Jones diagnostic criteria for ARF (2015).

Probable ARF: acute presentation which does not fulfil Jones diagnostic criteria for ARF (missing one major or one minor criterion or lacking evidence of preceding streptococcal infection) but ARF is still considered the most likely diagnosis.

Possible ARF: acute presentation which does not fulfil Jones diagnostic criteria for ARF (missing one major or one minor criterion or lacking evidence of preceding streptococcal infection) and ARF is considered uncertain but cannot be ruled out

Treatment of the acute rheumatic fever (Table 3)

- Antibiotic to eradicate GAS infection and prevent future reinfection,
- Anti-inflammatory therapy for symptomatic relief,
- Management of heart failure if present
- Other supportive treatment.

TABLE 3: Treatment of the Acute Rheumatic Fever

Antibiotic

All cases of ARF should receive Penicillin either injection or oral

Inj. Benzathine penicillin G (BPG) (1,200,000 units for wt ≥27kg
600,000 units for wt < 27 kg), Deep IM, single dose
OR

Phenoxymethyl Penicillin (Oral Penicillin V) (Children 250 mg PO TDS for 10 days, Adults 500 mg PO TDS for 10 days) OR

Erythromycin 10mg/kg/dose PO, BID for 10 days (if allergic to penicillin) OR

Azithromycin 10-12mg/kg/day PO, OD for 5 days (Max dose 500mg OD)

Aspirin

The usual dose of Aspirin is 50-60 mg/kg/day in 4 divided doses (QID) for 4 weeks. (Maximum up to 100mg/kg/day). Taper the dose once symptoms are resolved and the acute phase reactants (ESR, CRP) decrease. Total duration of treatment is 12 weeks or should be individualized depending upon the severity of illness.

Naproxen: If Aspirin intolerance is detected use Naproxen – Dose 10-20mg/kg/day in 2-3 divided doses

Steroids (prednisolone): Initiate after consultation with cardiologist if available

Prednisolone may be used in the following situations:

1. Patients who do not tolerate aspirin.
2. Patients who do not improve with aspirin.
3. Patients with moderate to severe Carditis (signs of heart failure)

Dose of prednisolone is 1-2mg/kg/day for 2 weeks then taper by 5mg every 2-3 days. Adult dose - max 80mg daily. While tapering steroid, overlap with aspirin (initial dose 50- 60mg/kg/day)

Continue **Aspirin** for 4-6 weeks after tapering off Prednisolone

- **Heart failure should be treated with**
- **Anti-failure medications** (e.g. Diuretics, ACEI/ARB, Aldosterone Antagonist, B-Blocker, Digoxin).
- **Limitation of physical activity and rest until symptoms get better.**

  Fluid and salt restriction

- **Chorea** Mild to moderate cases do not need medication. For severe cases Carbamazepine or Sodium Valproate or Haloperidol can be given.
- **Carbamazepine:** 3.5-10mg/kg/dose PO BID
- **Sodium Valproate:**7.5-10 mg/kg/dose PO BID
- **Haloperidol:** 0.25-0.5mg PO 2-3 times a day

How to prevent ARF?

There are two levels of prevention of ARF

Primary prevention of ARF:

Treatment of GAS throat infection to prevent the development of ARF (Table 1)

Secondary prevention of ARF:

Treatment aimed at prevention of recurrences of GAS throat, recurrences of ARF and progression of heart valve damage (Table 4)

TABLE 4: Secondary prevention of ARF

Drug	route	Dose	Frequency
Inj.BPG	IM	1,200,000 units for weight >27kg 600,000 units for weight ≤27 kg	3 weekly
PenicillinV (Phenoxymethyl Penicillin)	oral	250 mg (> 27Kg), 125mg (≤27 kg)	BID
Erythromycin (For those who are allergic to Penicillin). Contraindicated in liver disease.	oral	250 mg (> 27Kg), 125mg (≤27 kg)	BID

NB: Inj. BPG should be the drug of choice whenever feasible.

TABLE 5: Duration of secondary prevention

Category of Patient	Duration
No carditis (No valve damage)	5 years or 21years of age, whichever is longer
Minimum or Borderline RHD **(Stage A RHD)**	5 years or 21years of age, whichever is longer.
Mild Carditis OR healed Carditis OR mild RHD **(Stage B RHD)**	10 years or 25 years of age, whichever is longer
Moderate to severe disease without clinical complications **(Stage C RHD)**	Up to 40 years of age
RHD with clinical complications OR After heart valve surgery/PTMC **(Stage D RHD)**	Up to 40 years of age or lifelong

NB: Stages of RHD (A, B, C and D) are based on echocradiographic findings. (15)

Recommendations on Penicillin allergy testing and administration of injection BPG

Good practice statement (WHO 2024 guideline)

Penicillin allergy testing should not be used in patients who have no history of penicillin allergy and who are prescribed IM BPG for secondary prevention of RHD.

TABLE 6: Recommendations on Penicillin Skin Testing

1.	**Perform penicillin allergy skin test in the following situations:**
	Before first penicillin injection.
	NB: Perform Penicillin challenge test with oral penicillin in patients with H/O mild penicillin allergy. If no reaction Penicillin can be given.
2.	**Steps for penicillin skin test: (Intradermal Test)**
	a. Use 23-G needle.
	b. Clean the middle of forearm with spirit swab.
	c. Inject 0.1 ml of diluted BPG intradermal on the forearm. Circle this area and note the time on the fore arm
	d. Wait for 15 to 20 min.
	e. Look for local signs and symptoms of allergy (e.g., redness, inflammation, itching, erythema, swelling, blistering).
	f. If any of the local signs are present and if the swelling is >10 mm, the test is considered positive.
	g. In case of doubt repeat on the other arm with double strength test dose

TABLE 7: Recommendations on Safe BPG Delivery

1.	Take consent from the patient or his/her relative before the first penicillin injection.
2.	Record the brand name and batch number of the BPG.
3.	Reconstitute the BPG powder with 3.5 ml of sterile distilled water.
4.	Use 2 separate needles: 1 for pricking the vial and the other for injecting into the patient.
5.	Use 10 ml syringe and 21-G needle for deep intramuscular injection.
6.	Patient should lie down on trolley or bed on abdomen with head resting on pillow in a comfortable and relaxed position. In hospital settings, bed should be portable to rush the patient to the intensive care unit in case of emergency.
7.	Inject BPG deep intramuscularly in the upper outer quadrant of the buttock.
8.	Stay prepared for the treatment of possible anaphylaxis. The following medicines and instruments should be ready for emergency use:
	a. Adrenaline injection: One ampoule pre-loaded into the syringe.
	b. Dexamethasone
	c. Antihistamine injection.
	d. Intubation set /Ambu bag/suction machine

TABLE 8: Recommendations for Minimizing pain of BPG Injections

1.	Shake the powdered BPG vial after adding 3.5 ml of distilled water until the powder dissolves and an opaque, viscous, suspension is formed with a final volume of -5.0 ml. The penicillin crystal can easily pass through a 21-to23-G needle. If the crystals are attached to each other, they form large particles that get clogged inside the needle. To avoid this situation, reconstitution of the powder with 3.5 ml of distilled water rather than 3 ml is advised.
2.	Use 21-G taper cut needle for intramuscular injection.
3.	Properly select the injection site and apply finger pressure for 10 s.
4.	Stretch the skin at the injection site with the thumb and index finger.
5.	Prick deep IM and draw back the liquid medicine to test if the needle is in the right position. Then inject the liquid medicine at 900 with taper cut needle tip facing downward in vertical plane, which will cause minimum nerve end damage. Before injecting
6.	Never double prick with the same needle.
7.	Push the syringe slowly, applying sufficient pressure in a gradually increasing manner to allow the crystals in the viscous medicine to flow smoothly. It may take up to 1 min to push 5.0 ml of solution.
8.	Distract the attention of the patient away from the injection.
9.	Maintain the Injection delivery room temperature below 300C. In hot air and moist skin, the injections are more painful.
10.	Apply ice pack in case of pain immediately after injection.
11.	mix 0.5 to 1.0 ml of 1% lignocaine with the BPG solution for reducing pain if all other techniques fail.

Signs, Symptoms and Management of anaphylactic reaction

Signs and symptoms (develop within 30 sec to 2 hours after administration of Injection penicillin)

1. Itching and rashes all over the body
2. Dizziness or Fainting
3. Fast heart rate (Tachycardia)
4. Low BP
5. Wheezing with Difficulty in breathing

Management

a. Injection Adrenaline (1:1000): 0.5 ml IM or S/C. Repeat after 10 minutes if needed. (Pediatric dose 0.01mg/kg IM)

b. Injection Dexamethasone 8 mg IM (Pediatric dose 0.15mg/kg/dose)

d. Injection Antihistamine IM

e. Oxygen

f. IV 5% dextrose saline

Action protocol on diagnosis, management and prevention of Acute Rheumatic fever

Action 1: Ask

- Ask about the symptoms for Rheumatic fever (One or more Joints pain, difficulty in walking or moving the hands, swelling of the large joints with migrating nature, jerky movements of limbs and facial muscles and Fever)
- Ask if there were similar symptoms in the past (If yes think about recurrence of ARF)
- Ask about the past history of bacterial tonsillopharyngitis.
- Ask how often did the throat infection occur.
- Ask about shortness of breath on exertion, palpitation and chest pain

Action 2: Assess (Examine the patient)

- Examine the joints of hand and legs (shoulder, elbow, wrist, hip, knee, ankle)
- Look for signs of inflammation of one or more large joints (painful, swelling, hot and redness) **(Monoarthritis or Polyarthritis)**
- Multiple joints pain **(polyarthralgia)** or single joint pain **(monoarthralgia)**
- Auscultate for Heart murmur **(Carditis)**
- Look for rashes in the body especially the trunk and abdomen (non-itching rashes with red circular margins) **(Erythema marginatum)**
- Look for small pea size nodules below skin of elbows, wrists, knees and other bony prominences of the limbs) **(Subcutaneous nodules)**
- Look if there is involuntary jerky movements of the hands, legs, head and facial muscles**. (Chorea)**

Action 3: Estimate

- Record the temperature (>100º F is suggestive)
- Estimate ESR (>30mm/h suggestive) and CRP > 3 mg/dl (positive is suggestive)
- Perform ECG and estimate the PR interval (prolonged PR more than 0.20 sec. is suggestive)
- Estimate the ASOT (positive or more than 200 units is suggestive)
- Use modified Jones criteria 2015 for establishing the diagnosis of ARF (**Table2**)

Action 4: Treat or refer if necessary

All cases of suspected or confirmed ARF should undergo Echocardiography test if available. If not available then the patient should be referred to the nearest health facility where echocardiography and cardiologist or physician is available. If that is going to be delayed, initiate the treatment as per guidelines **(Table 3)** and then refer to higher center.

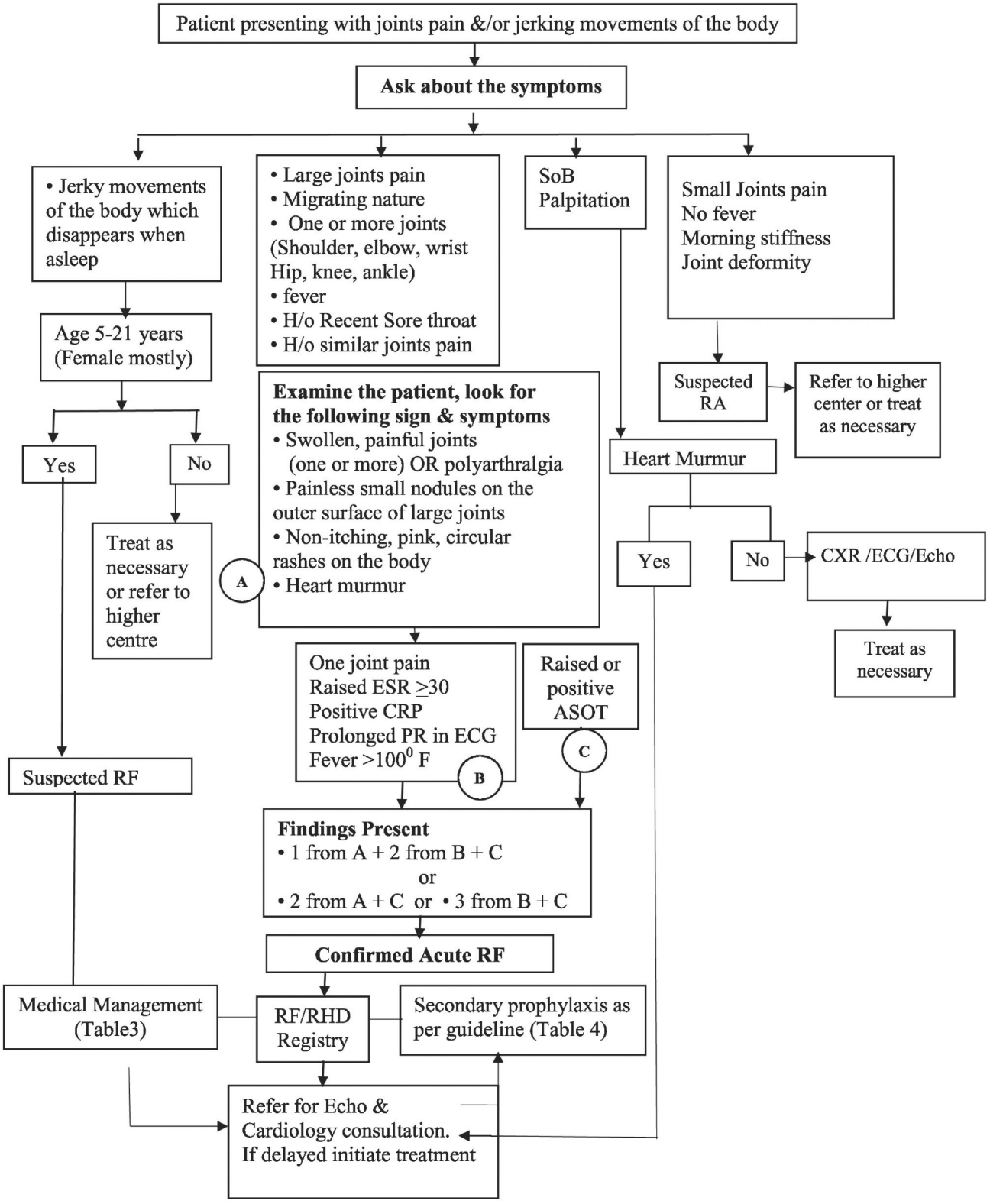
Action 5: Counsel

- Explain the patient or the parents about the seriousness of illness and the importance of early treatment
- Explain about the need and importance of secondary prophylaxis (**Table 4**)

Action 6: Registry and follow up of the patients with confirmed diagnosis of ARF

- Register the case and provide follow up for injection BPG every 3 weeks or oral penicillin. In case of migration of the patient to another location, refer the patient to respective location for follow up.
- If patient is allergic to penicillin follow up monthly during refill of Erythromycin

Algorithm for Suspected Acute Rheumatic Fever (ARF)



CHAPTER - 4

DIAGNOSIS, MANAGEMENT AND PREVENTION OF RHEUMATIC HEART DISEASE (RHD)

Rheumatic Heart Disease

Introduction

- Rheumatic heart disease is a condition of heart valve damage resulting from repeated episodes of ARF.
- Rheumatic heart disease is a condition of heart valve damage resulting from repeated episodes of ARF.
- Early diagnosis of RHD is very important so that secondary prophylaxis can be started as soon as possible to prevent the progression of the heart valve damage.
- Echocardiography is an important test to confirm the diagnosis of RHD.
- The mitral valve is affected in over 90% of cases of RHD. The next most commonly affected valve is the Aortic valve.

Types of RHD

RHD can be divided broadly into clinical and subclinical.

Clinical RHD: Clinical RHD are those, which are symptomatic and have murmur on auscultation.

Subclinical RHD: Subclinical RHD are diagnosed only with echocardiography. On auscultation they do not present with heart murmur. These are silent cases and are in very large number in the community (10 times more than clinical RHD). Echocardiographic screening is useful in the diagnosis of such cases.

Echocardiography describes **Four stages of RHD based on severity of valve damage**

a) Stage A (Minimum RHD) (Borderline RHD)

b) Stage B (Mild RHD), includes Mild MR, Mild AR

c) Stage C (Advanced RHD without clinical complications), includes mod-severe MR, AR and any MS, AS)

d) Stage D (Advanced RHD with clinical complications)

Staging of rheumatic heart disease detected by echocardiography (15)

Stage A: minimal echocardiographic criteria for RHD. Applies only to individuals aged d”20 years old Clinical risk: might be at risk of valvular heart disease progression. Echocardiographic features: the presence of mild MR or AR without morphological features

Stage B: mild RHD. Can apply to any age. Clinical risk: at moderate or high risk of progression and at risk of developing symptoms of valvular heart disease. Echocardiographic features: evidence of mild valvular regurgitation plus at least one morphological feature in individuals aged d”20 years and at least two morphological features in individuals aged >20 years; or mild regurgitation in both mitral and aortic valves

Stage C: advanced RHD at risk of clinical complications. Can apply to any age. Clinical risk: at high risk of developing clinical complications that require medical or surgical intervention. Echocardiographic features: moderate or severe MR, moderate or severe AR, any MS or AS, pulmonary hypertension and decreased LV systolic function

Stage D: advanced RHD with clinical complications. Can apply to any age. Clinical risk: established clinical complications include cardiac surgery, heart failure, arrhythmia, stroke and infective endocarditis.

Echocardiographic features: moderate or severe MR, moderate or severe AR, any MS or AS, pulmonary hypertension and decreased LV systolic function

Symptoms of RHD

The symptoms of RHD depend on the valve lesion and its severity. Symptoms of RHD may not show for many years until valve damage becomes severe.

Symptoms of RHD are the symptoms of early heart failure. They are:

- Breathlessness on exertion
- Feeling tired
- General weakness

As heart failure progresses, other symptoms may develop including:

- Orthopnea (breathlessness on lying down)
- Paroxysmal nocturnal dyspnea (waking at night with shortness of breath)
- Peripheral oedema
- Palpitations (feeling of irregular heartbeats).
- Chest pain (People with aortic valve disease may experience chest pain)
- Syncope

How to diagnose RHD?

Physical Examination

- Careful auscultation should be undertaken and suspicious murmurs should be further evaluated for confirmation of RHD by echocardiography.
- With physical examination RHD can only be suspected.
- Echocardiography test is necessary to confirm the diagnosis of RHD.

Laboratory tests

ECG is essential to determine the rhythm and heart rate.

Chest X-Ray helps to assess the size of the heart chambers and to detect pulmonary congestion.

Echocardiography is confirmatory test for RHD. 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; help determine its severity, pulmonary pressure and left ventricular function. Regular echocardiography helps to detect evidence of progression of valve lesions over time and to assess heart function before surgery.

Complications of RHD

RHD if not managed properly may lead to following complications

- Arrhythmia (mostly AF)
- Heart failure
- Infective endocarditis
- Stroke (thromboembolic events)
- Pulmonary hypertension

Table 9: Common Medicines used in the management of ARF and RHD

Medication	Indication	Dose/Route/Frequency/Duration	Contraindications
Furosemide	Heart failure	Adult: 20mg once daily Pediatric: 1-2 mg/kg every 12 hours PO or IV **Max dose: 240mg BD**	Low Na, K Dehydration Hypotension
Spironolactone	Heart failure	Adult:12.5 to 25mg once daily Pediatric dose: 1-3.5 mg/kg/day PO in single **Max dose: 50mg OD**	High K Renal failure
Torsemide	Heart failure	Adult: 10mg OD **(Max dose 200mg OD)** Pediatric: safety and efficacy not established	Low Na, K Dehydration, Hypotension
Warfarin	AF	Usually started with 2 to 5 mg per day (0.1–0.2 mg/kg/dose) and the dose is adjusted to target INR of 2-3	Active bleeding, bleeding, disorders, pregnancy
Digoxin (Oral)	Tachycardia, AF, CCF	0.125-0.5mg/day OD (5-6 days/wk) OR Pediatric dose:5-10 mcg/kg/day (5 days/wk)	Bradycardia, heart blocks, Renal failure
Metoprolol Succinate	Tachycardia, CCF	Adult dose:25mg OD **(Max dose: 200mg OD)** Pediatric dose not established	Bradycardia, Heart blocks, Hypotension Bronchial asthma
Enalapril	Heart failure	Adult dose :2.5 mg BD (**Max dose:10mg BD)** Pediatric dose: 0.1 mg/kg/d PO divided OD/BID, not to exceed 0.5 mg/kg/d	Pregnancy, High K, Renal failure Hypotension
Losartan	Heart failure	Adult: 25mg daily **(Max dose: 50mg BD)** Pediatric: 0.75-1.4 mg/kg/day	Same as Enalapril
Gentamycin, Cetriaxone and /or Vancomycin	Infective Endocarditis	Ceftriaxone: 2 g IV BD (adult); 100 mg/kg/day IV in 2 equally divided doses (pediatric) Gentamycin: 3 mg/kg/day IV in 1 dose (adult) 3 mg/kg/day IV in 3 divided doses (pediatric)Vancomycin: 30 mg/kg/day IV in 2 doses (adult) 40 mg/kg/day IV in 2 or 3 equally divided doses (pediatric)	

NB: RHD with complications (heart failure, AF, Stroke, Infective endocarditis) should be referred to higher center at the earliest possible. If referral is delayed then initiation of treatment is advised.

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(1 - doses of antibiotics in Infective Endocarditis, 2 and 3 - doses of heart failure drugs in pediatrics)

Management of RHD (Table 9)

1. **Medical management**

   Heart failure-Diuretics (Frusemide), Aldosterone antagonist (Spironolactone), ACEI (Enalapril)/ ARB (Losartan), Digoxin, B Blocker (if not contraindicated)

   Atrial fibrillation (AF) - beta blocker or Digoxin (for rate control)

   Anti-coagulant - Warfarin or NOAC (Rivaroxaban, Apixaban) is indicated if AF is present.

   Warfarin should be used in AF with Mod-Sev MS or Mechanical Prosthetic Valve. It can be used in any conditions of ARF/RHD with AF. Dose- 1-10mg OD. Loading dose in adults: 5mg OD, in children and elderly: 2-3 mg OD. Maintain INR at 2-3.

   NOAC can be given in AF with Pure MR or post MV Repair without residual significant stenosis or Aortic valve disease or tricuspid valve disease.

   NB: PT/INR monitoring for NOAC is not needed. However, the use of NOAC should not be encouraged because they are expensive and the antidote is not readily available. Anticoagulant should be used cautiously because of possible bleeding risk.

   Infective Endocarditis- IV Antibiotics (Ceftriaxone, Gentamycin and/or Vancomycin)

   LA thrombus (clot) – Warfarin

2. **Surgical /interventional management:**

   In case of Advanced RHD

   PTMC/BMV-If Mitral valve is suitable for PTMC/BMV

   Valve repair/replacement – Valvular damage not suitable for PTMC/BMV

Strategies for treatment of AF

Strategies	Drugs
Rate control – for hemodynamically stable patients with chronic AF and fast AV conduction	Digoxin, calcium channel blockers (Verapamil, Diltiazem), beta-blockersc (Metoprolol, Bisoprolol, Atenolol)
Rhythm control – for hemodynamically unstable patients with AF of recent onset	Electrical cardioversion Pharmacological cardioversion-Inj. Amiodarone IV infusion (5-7mg/kg over 1-2 hours, followed by 50gm/hours to a maximum of 1gm over 24 hrs. Dilute in 5% glucose)
Anticoagulation – for all patients	Warfarin to achieve INR of 2-3

Source: Consensus Statement of Cardiac Society of Nepal on Diagnosis, Management and Prevention of Acute Rheumatic Fever and Rheumatic Heart Disease in Nepal. *Nepalese Heart Journal 2022; Vol 20(2), 37-48*

Recommendations for follow-up of RHD patients

Moderate- Severe RHD with complications **(Stage D)** AND Pregnant women with RHD (of any severity)	Echocardiogram and Specialist review: at least 6 monthly
Moderate -Severe RHD without complications **(Stage C)**	Echocardiogram and specialist review: Yearly
Mild RHD **(Stage B)**	Echocardiogram and specialist review: Yearly
Borderline or minimum RHD **(Stage A)** AND ARF without valve damage	Echocardiogram and specialist review: Yearly after diagnosis. At 1 year, 3 years and 5 years post cessation of secondary prophylaxis

RHD and Pregnancy

Pregnancy causes stress to the heart and may make any existing valve problem worse. The cardiovascular changes which occur during pregnancy with RHD may threaten the health of the woman and the fetus. Changes that occur during pregnancy are:

- Increase in heart rate and blood volume
- Decrease in systemic and pulmonary resistance
- Increase in cardiac output.

  Women with RHD are at high risk of complications.
  Women with known RHD should be fully assessed before pregnancy.
  Women at high risk may be counseled to avoid pregnancy (e.g. severe pulmonary hypertension).
  When pregnancy occurs, management depends on the type and severity of heart valve disease.
  Pregnant woman with RHD should be assessed by a medical Physician/Cardiologist as early as possible so that a coordinated pregnancy management and follow-up can be planned.

Management includes:

- Restriction in physical activity and added salt intake
- Appropriate secondary prophylaxis

  Special attention should be given to women with high risk RHD including women with
  - Mitral and/or aortic stenosis
  - Atrial fibrillation
  - Prosthetic heart valves
  - Those receiving anticoagulant therapy

RHD in pregnancy - risk levels

Level I: low risk of maternal mortality, low to moderate risk of morbidity (e.g. mild RHD with no mitral stenosis).

Level II: elevated risk of maternal mortality or moderately increased risk of morbidity (e.g. bioprosthetic valve or mild mitral stenosis).

Level III: further elevated risk of maternal mortality or severe morbidity (e.g. mechanical heart valve, severe asymptomatic mitral / aortic regurgitation or severe asymptomatic aortic stenosis or moderate mitral stenosis.

Level IV: extremely high risk of maternal mortality or severe morbidity (e.g. severe mitral stenosis or valve disease with pulmonary hypertension).

Key points in the management of pregnancy in women with RHD

Predictors of increased maternal and fetal risk	Decreased LV systolic function Significant aortic and mitral stenosis Moderate or severe pulmonary hypertension Heart failure symptoms before pregnancy Mechanical valve prostheses Atrial fibrillation
Cardiac assessment	Early echocardiography to assess valves and LV function Plan multidisciplinary management
Mitral/aortic regurgitation	Usually well tolerated Treat medically with diuretics, vasodilators (no ACE inhibitors/angiotensin II receptor blockers)
Mitral stenosis (MS)	Mild to moderate MS - manage medically. Moderate to severe MS (MVA<1.5 cm^2) - consider PTMC during late second trimester, if patient remains symptomatic and PASP >50 mmHgBeta-blockers or digoxin for rate control of AF
Aortic stenosis (AS)	Mild to moderate AS – well tolerated. Manage medically. Diuretics for heart failure Consider PTAV if severe symptoms Beta-blockers or digoxin for rate control of AF. Avoid cardiac surgery, as high risk of fetal loss
Mechanical /prosthetic valves and anticoagulation in pregnancy Choice of 3 antithrombotic regimens	High maternal and fetal risk Risk of warfarin embryopathy in first trimester Embryopathy may be avoided if warfarin dose ≤5 mg 1. LMWH throughout pregnancy 2. Warfarin throughout pregnancy, if can keep dose <5 mg (INR 2-3), change to LMWH or unfractionated heparin at 36 weeks 3. LMWH until 13 weeks, and then warfarin and Aspirin until 36 weeks; change to LMWH or UFH until labour.
Labour	Haemodynamic monitoring: non-invasive, if mild to moderate valve disease. Antibiotic prophylaxis, if prolonged labour and/or ruptured membranes.Aim for short second stage and multidisciplinary management approach.If patient is symptomatic refer the case to higher center where cardiologist is available

ACE- angiotensin-Converting enzyme; INR- internal normalized ratio; LMWH- low-molecular weight heparin; LV- left ventricle; MVA- mitral valve area; PASP- pulmonary artery systolic pressure; PTMC- percutaneous transluminal mitral commissurotomy; PTAV- percutaneous transluminal aortic vavuloplasty; UFH- unfractionated heparin.

Prevention of RHD

1 Primordial Prevention:

- The measure taken to prevent the development of GAS throat infection.
- Improvement in the socio-economic condition, sanitation, standard of living, awareness raising, access to healthcare etc.

2 Primary Prevention:

- Early treatment of GAS throat with adequate antibiotic.
- This measure can prevent the development of ARF.

3 Secondary Prevention:

- Prevent the recurrence of GAS infection and ARF by administering injection BPG every 3 weeks. Keep registry of RF/RHD patients.
- This measure further prevents the progression of heart valve damage.

4 Tertiary Prevention:

- Prevent the life-threatening complications of RHD like heart failure, stroke, infective endocarditis etc.
- This is done through medical and/or surgical interventions.

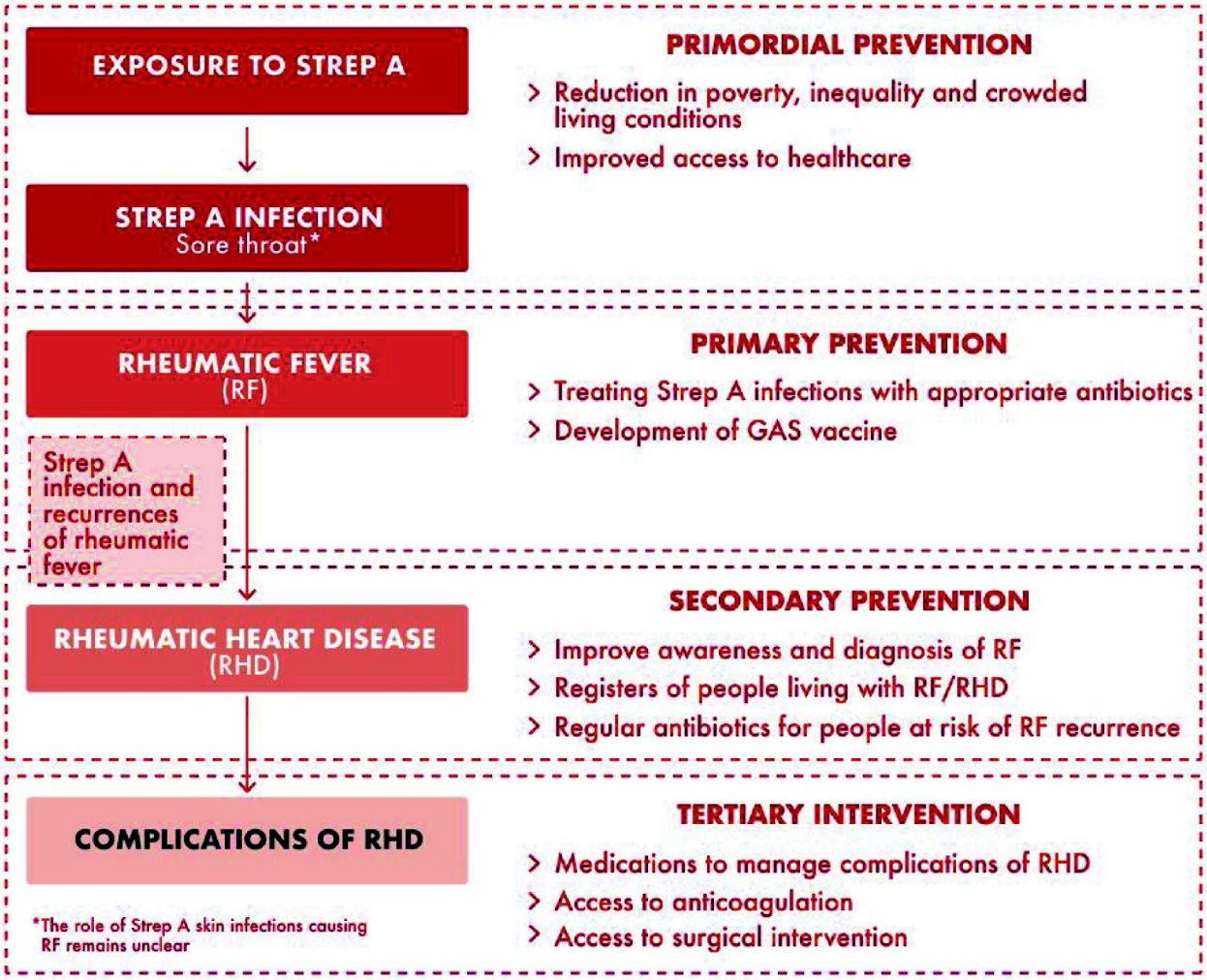


Figure 1: Prevention strategies at different level of intervention

RHD prevention and control program

To control RHD, a comprehensive RHD control program is needed which should be implemented at the national level by the government. World Heart Federation (WHF) has described a framework of a comprehensive RHD program which consists of 25 components. According to the WHO the following components are the basic pillars for a successful RHD control program:

1. The program should be register based, preferably a computer register (**Registry of RF/RHD cases**)
2. It should be integrated into the existing health care system (**Diagonal approach**)
3. There should be a commitment for funding by the government. (**Sustainability**)

Necessary components for initiating RHD control program

1. Awareness raising
2. Primary prevention through all health facilities
3. Screening for early detection of RF/RHD
4. RF/RHD Registry
5. Secondary prophylaxis delivery (Inj. BPG or oral Penicillin) from all levels of health facilities
6. Availability of Inj. BPG and oral Penicillin
7. Surgery/PTMC facility or needy cases
8. PT/INR testing facility at all levels hospitals
9. Availability of Warfaring
10. Training

Responsibilities of health facilities

Responsibilities of Basic Health Services Centres (BHS)

- Diagnose and Treat BTP
- Keep RF/RHD Registers
- provide oral penicillin and follow up care
- Refer when needed

Responsibilities of Basic Hospitals (BH)

- Diagnose and Treat BTP
- Suspect/confirm ARF
- Keep RF/RHD Registers
- Delivery of Inj. BPG and oral penicillin,
- Follow up care
- Refer when needed

Responsibilities of District Hospitals (DH)

- Diagnose and treat BTP and ARF
- Diagnose RHD if facilities available (Echocardiography divice & Cardiologist or Echo Trained Physician)

- Keep RF/RHD Registers
- Delivery of Inj. BPG and oral penicillin,
- Follow up care
- Refer when needed

Program implementation plan

Phase 1

1. Make available oral penicillin, Amoxycillin, Erythromycin (BHSm BH and DH), Inj BPG (at BH, and DH)
2. Make avialable laboratory test facilities for ESR, ASOT, CRP, PT/INR and ECG (at BH, DH)
3. Keep RF/RHD Register (Paper and Computer) at all levels of health services (BHS, BH, DH, PH, FH)
4. Register confirmed cases of RF/RHD and Provide BPG every 3 weeks or oral penicillin free of cost (BHS will provide Penicillin V; BH and DH will provide BPG and Penicillin V as needed)
5. Continue and expand the PEN Plus clinics for RHD care at District hospitals
6. Publish a program manual with all information on how to conduct the program
7. Orientation training

Phase 2

1. Training of nurses or paramedics in RHD focused echocardiography and engaging them in RHD screening camps
2. Publish training manual for nurses in RHD focused echocardiography
3. Ensure availability of medicines for treatment of RHD with complications (Warfarin, Frasemide) ACEI/ARB, B-Blocker, Digoxin etc. (at DH, PH and FH)

(BHS-Basic health service, **BH-** Basic hospital, **DH-** District hospital, **PF**- Provincial hospital, **FH**-Federal hospital)

Action protocol on diagnosis, management prevention of RHD

Action 1: Ask

- Ask about the symptoms for Rheumatic heart disease
  - SOB on exertion,
  - Palpitation,
  - Chest pain,
  - Haemoptysis
  - Paroxysmal nocturnal dyspnoea (awakening at mid night due to dyspnoea)
  - Swelling of legs
- Ask about the past history of bacterial tonsillopharyngitis.
- Ask about the past history of joints pain, swelling and fever (ARF)

Action 2: Assess (Examine the patient in supine position)

- Look of raised JVP
- Look for swelling of legs
- Feel the pulse for arrhythmias (AF)
- Auscultate to detect any Heart murmurs

Action 3: Estimate

Suspect RHD if few of the following signs, symptoms and abnormal tests are present

- Age 5-45years
- Heart murmur
- SOB on exertion
- Swelling of legs
- Irregular pulse
- Past history of multiple joints pain with or without swelling and fever
- Abnormal ECG
- Cardiomegaly in Chest X-Ray PA

Action 4: Treat or refer if necessary

All cases of suspected RHD should be immediately referred to the nearest health facility where echocardiography and cardiologist or physician is available. If that is going to be delayed initiate treatment as indicated.

Action 5: Counsel

Explain the patient or the parents about the seriousness of illness and the importance of early treatment and long- term secondary prophylaxis

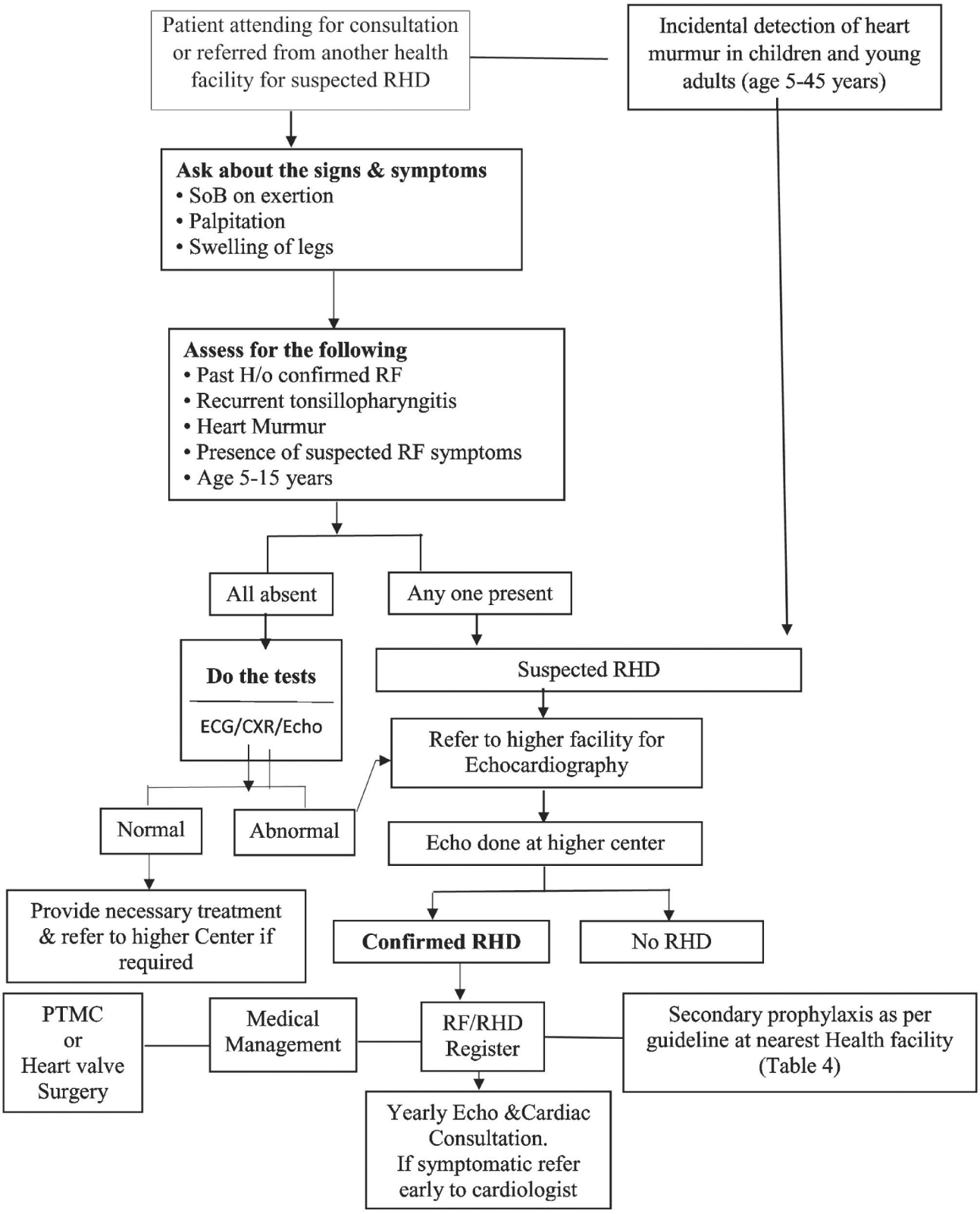
Action 6: Registry and follow up of the patients with confirmed diagnosis

Register the patient into the RHD register

Provide follow up for secondary prophylaxis with inj. BPG, oral penicillin or Erythromycin.

In case of migration of the patient to another location, refer the patient to respective location for follow up care.

Algorithm for Suspected Rheumatic Heart Disease (RHD)



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