Acute Rheumatic Fever & Rheumatic Heart Disease

Diagnosis, Management and Prevention

Physician Training Module

Contents

1. Introduction
2. Primary Prevention:

   Diagnosis and management of Sore Throat

3. Secondary Prevention:

   Diagnosis and management of Acute Rheumatic Fever

4. Safe Administration of Benzathine Penicillin
5. RHD and Control Programs

Course Objectives

To have good knowledge on:

1. Diagnosis and management of bacterial pharyngitis
2. Diagnosis and management of acute rheumatic fever
3. Safe administration of Benzathine penicillin
4. Rheumatic Heart Disease (RHD) and its control program

Pre- Test

Chose the best answer:

1. The best way to diagnose Bacterial Pharyngitis (BPh) in patients 3-18 years of age in endemic areas is by:

A. Clinical findings

B. Throat culture and rapid antigen test

C.High leukocyte count

D. High ESR

2. The best way to treat BPh in patients 3-18 years is:

A. Oral penicillin or cephalosporin for 5-7 days

B.IM Benzathine penicillin (one injection)

C. Azithromycin for 3 days

D. Erythomycin for 7 days

3. Primary prevention of acute rheumatic fever (ARF) means:

A. Prompt diagnosis and treatment of BPh

B. Giving 3 weekly penicillin

C. Diagnosis and treatment of ARF

D.Management of rheumatic Heart Disease

4. Which one of the following is a major criteria of ARF:

A.Fever B.High ESR C.Monoarthritis D.High ASO

5. Which one of the following is a minor criteria of RF:

A.Monoarthritis B. Polyarthralgia C.Skin nodules D.Monoarthrlagia

6. Which one of the following is not a feature of rheumatic carditis:

A.Pansystolic apical murmur B.Early diastolic murmur with large volume pulse

C.Heart failure without murmurs D.Large volume pulse with Corigan sign

7. Diagnosis of ARF in new patients includes:

A. One major criteria plus high ASO titre B. 2 minor criteria plus high ASO titre C. Subclinical carditis plus high ASO D. Two major plus high ASO

8. Diagnosis of RECURRENCE OF ARF is made with :

A. Two minor criteria plus high ASO titre B. Is made in the same way of new episode ARF.

C. Carditis without other criteria D. Fever with high ASO

9. Treatment of ARF includes all the following <u>except</u> :

A. High dose Aspirin B.Bed rest C.Benzathin penicillin D.Steroids in all patients with carditis.

10. Secondary prophylaxis of ARF means:

A. Giving 3 weekly benzathine penicillin to prevent recurrence

B. Giving benzathine penicillin to all patients with high ASO titre

C. Aspirin in a dose of 75 mg per kg per day for 4 weeks

D. Single injection of benzathine Penicillin for sore throat.

11.When giving benzathine penicillin:

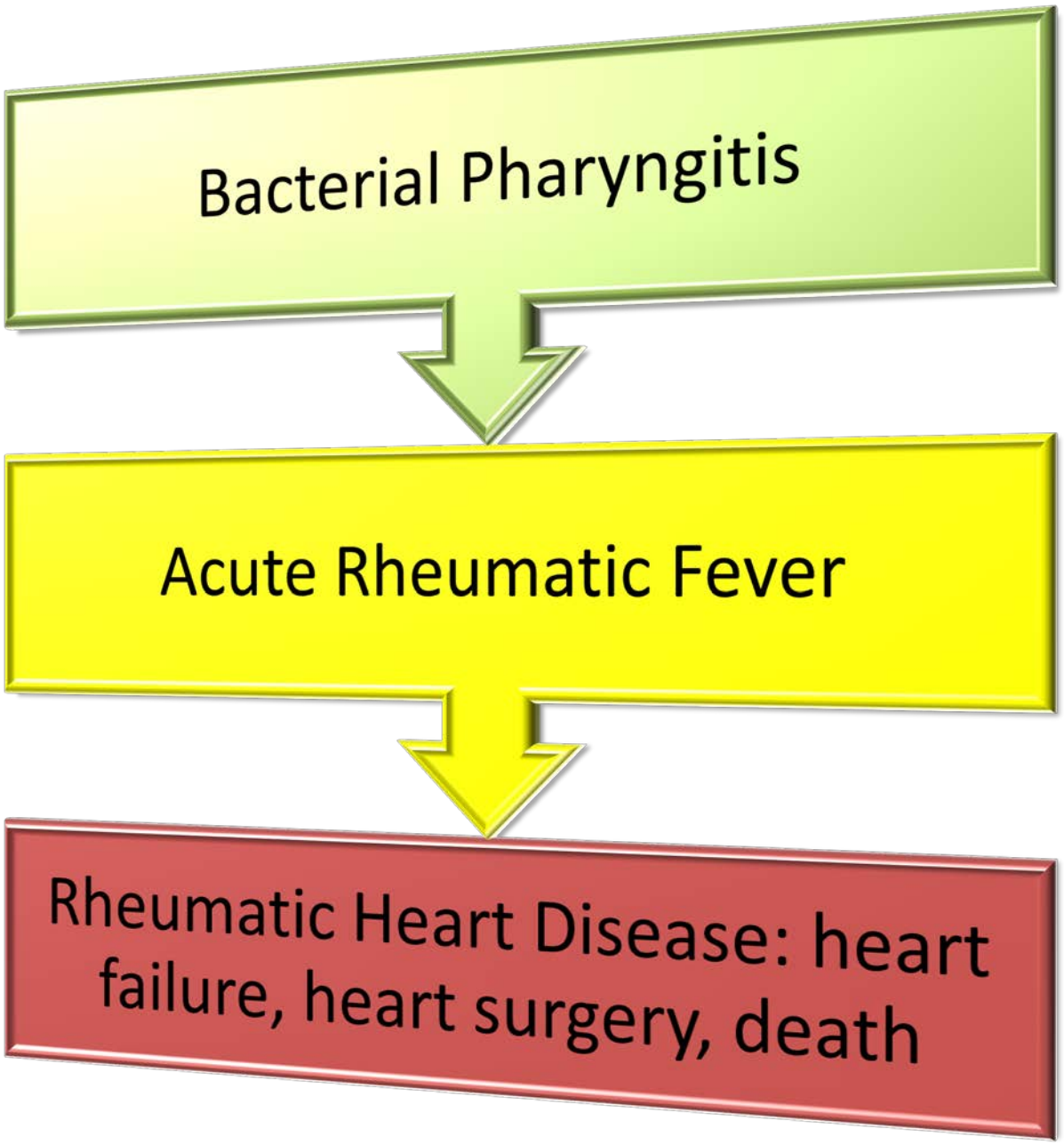
A. Skin test should NOT be done B. You need to have adrenalin

C. Lidocaine is used to decrese pain D.All the above

Introduction

RHD

- RHD is inflammation of heart valves that follows infection with Group A beta hemolytic streptococcus, commonly pharyngitis.


Bacterial Pharyngitis
Acute Rheumatic Fever
Rheumatic Heart Disease: heart failure, heart surgery, death

RHD is a Global Health Problem

- 18 Million people affected, mainly from developing countries
- The most common cause of acquired heart disease in the young

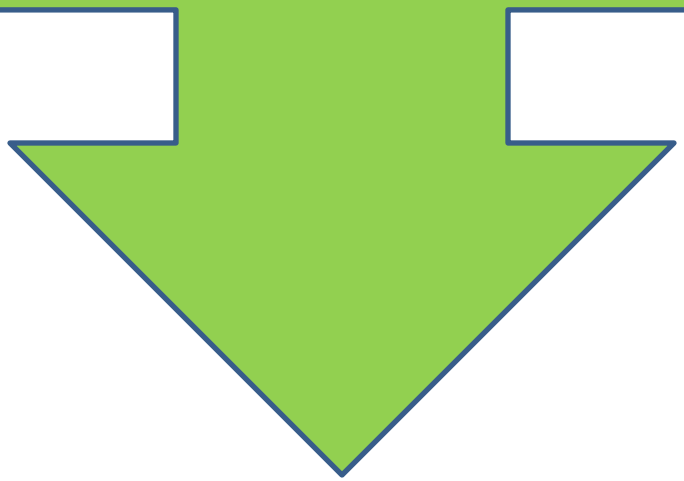
Carapetis, J. R., Steer, A. C., Mulholland, E. K. & Weber, M. The global burden of group A streptococcal diseases. *Lancet Infect. Dis 2005;*5: 685–694 .

Painful Facts

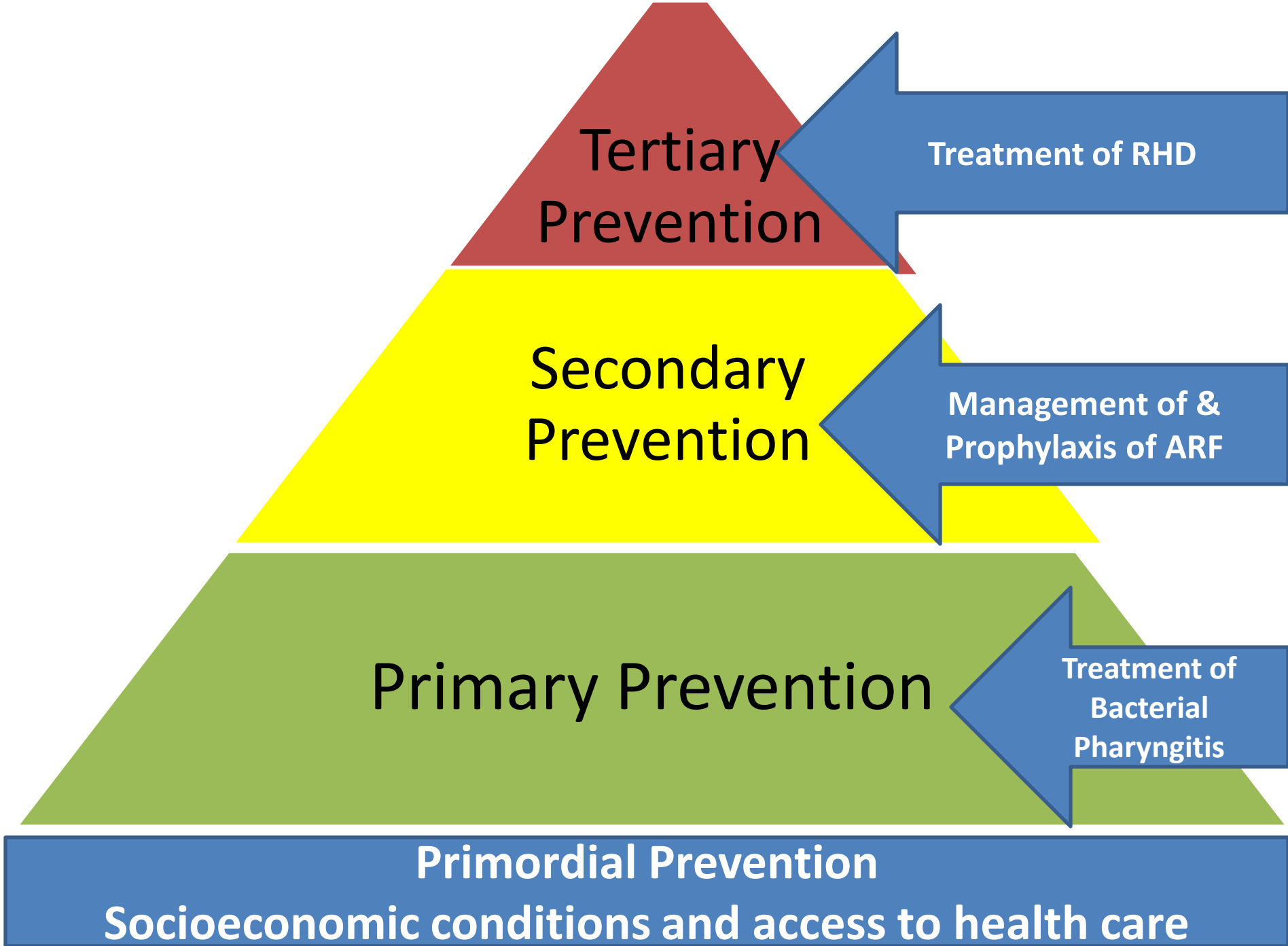
- Most patients in developing countries present with severe disease due to late diagnosis
- Only a minority have access to surgery
- Valve replacement: costs 4-10 000 USD and not readily available
- Poor long term outcome after surgery

Eynas Khalid, Sulafa Ali et al.Clinical and echocardiographic features of 370 children with rheumatic heart disease seen in Khartoum. Sudan Med J 2014;50:151-54

RHD is preventable with early diagnosis and management of strep pharyngitis and ARF



The Levels of RHD Prevention


Tertiary Prevention
Treatment of RHD
Secondary Prevention
Management of & Prophylaxis of ARF
Primary Prevention
Treatment of Bacterial Pharyngitis
Primordial Prevention
Socioeconomic conditions and access to health care

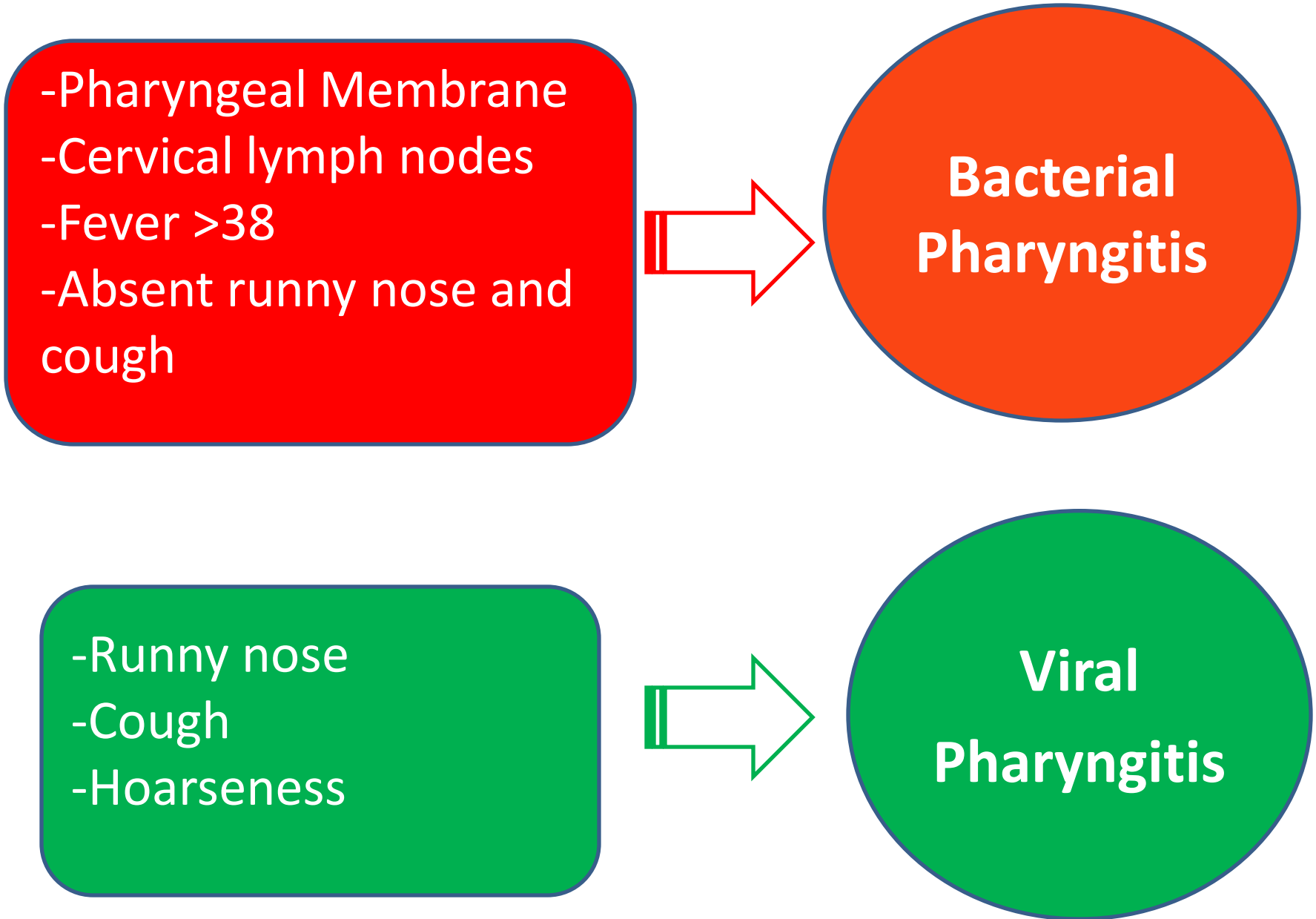
Primary Prevention

Definition of Primary Prevention

“ Prompt diagnosis and treatment of Streptococcal pharyngitis”

Sore Throat

- Mostly viral
- Bacterial about 30% caused by Group A streptococcus (GAS)
- Diagnosis of Bacterial Pharynges is clinical


-Pharyngeal Membrane
-Cervical lymph nodes
-Fever >38
-Absent runny nose and cough
Bacterial Pharyngitis
-Runny nose
-Cough
-Hoarseness
Viral Pharyngitis

How can we diagnose bacterial pharyngitis?

- Diagnosis is clinical
- Not practical to do throat cultures or rapid antigen test in limited resource settings.

ASO Titer:
NO role in acute Pharyngitis

Titers increase only 7 to 14 days ***after*** the onset of infection and remain high for weeks

Michael R. WesselsN Engl J Med 2011;364:648-55.

Clinical Algorithms for Bacterial Pharyngitis

WHO: IMCI Program uses 3 Points

• fever **OR** Sore throat AND **Two of the following :** • Red (congested) throat • White or yellow exudate on the throat or tonsils. • Enlarged tender lymph node(s) on the front of the neck.	**STREPTOCOCCAL SORE THROAT**	▯ ***Give benzathine penicillin.*** ▯ Soothe the throat with a safe remedy. ▯ Give paracetamol for pain. ▯ Advise mother when to return immediately. ▯ Follow up in 5 days if not improving.

WHO Algorithm:3 POINTS

- 12% sensitive and 94% specific
- Missed 88% of children with positive cultures

Rimoin AW, Hamza HS, Vince A, Kumar R, Walker CF, Chitale RA, daCunha ALA, Qazi S, Steinhoff MC.Evaluation of the WHO clinical decision rule for streptococcal pharyngitis. Arch Dis Child. 2005;90:1066–1070.

2 Point Algorithms

Two of the following (sore throat, fever, pharyngeal erythema and pharyngeal exudates)

Sensitivity of 80% and specificity of 40%.

Steinhoff MC, Abd el Khalek MK, Khallaf N, Hamza HS, el Ayadi A, Orabi A, Fouad H, Kamel M Effectiveness of clinical guidelines for the presumptive treatment of streptococcal pharyngitis in Egyptian children. **Lancet**. 1997;27:918-21.

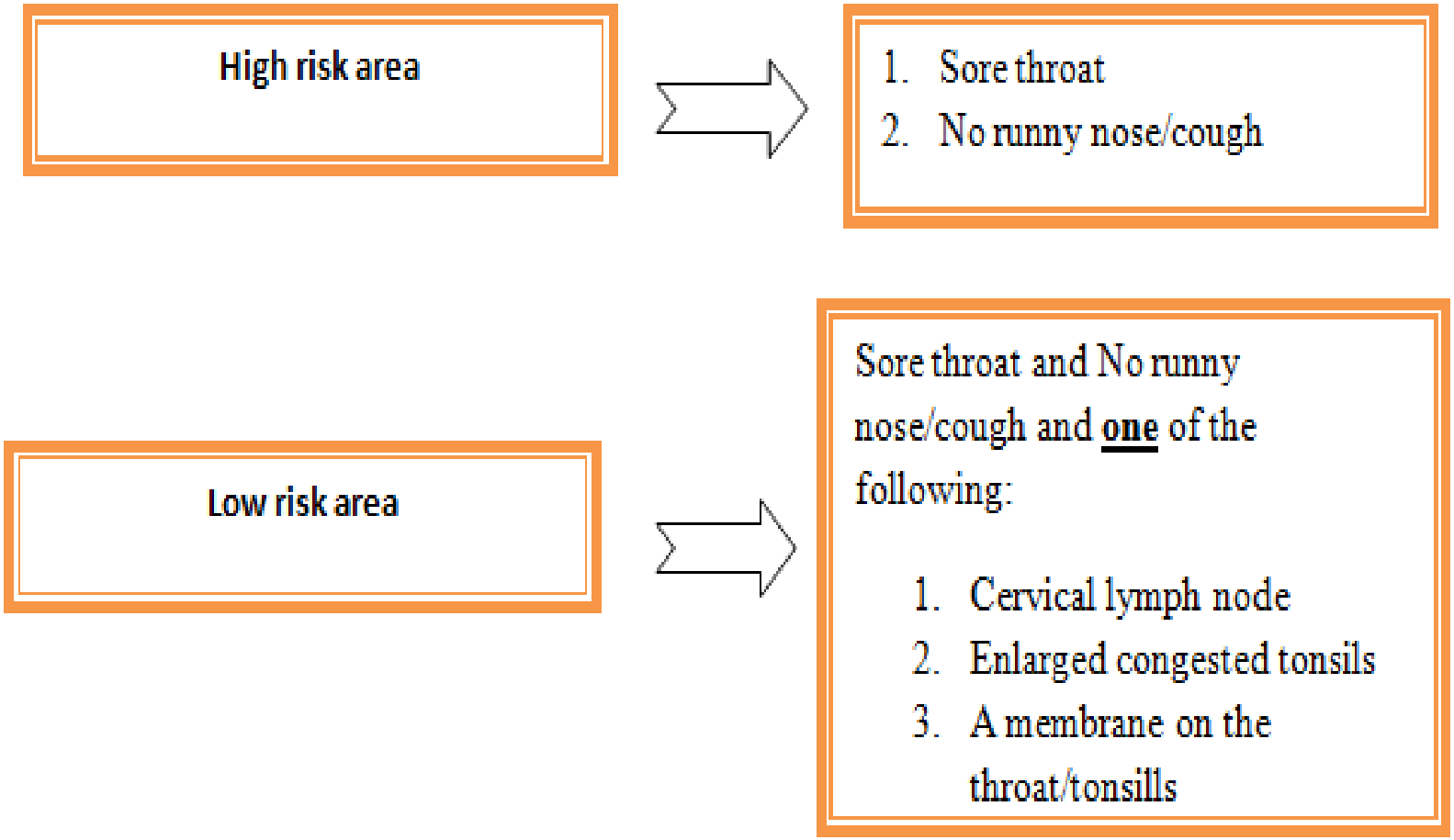
Sahin F, Ulukol B, Aysev D, Suskan E. The validity of diagnostic criteria for streptococca l pharyngitis in Integrated Management of Childhood Illness (IMCI) Guidelines. J Trop Pediatr. 2003;49:377–379.

The Mosi-o-Tunya Call to Action

- Treat all children with sore throat with BPG

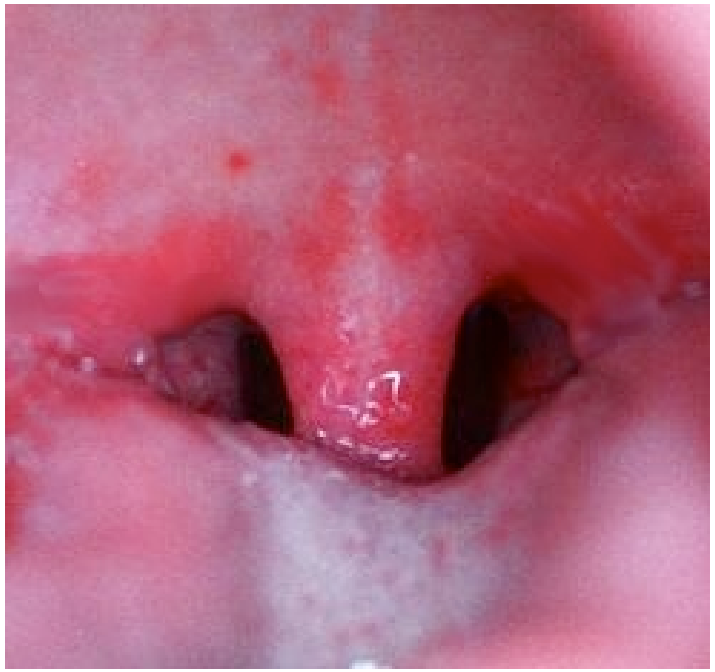
Bongani M Mayosi, Habib Gamra, Jean-Marie Dangou,Joseph Kasonde, for the 2nd All-Africa Workshop on Rheumatic Fever and Rheumatic Heart Disease participantsRheumatic heart disease in Africa: the Mosi-o-Tunya call to action.The **Lancet2014**;2:e438-9

Proposed Clinical Algorithm
In patients 3-18 years



Important

- Bacterial pharyngitis can affect tonsils (tonsillitis) or peritonsilar area
- Tonsillectomy may decrease but does not prevent ARF



Treatment of Bacterial Pharyngitis

- **One injection Benzathine penicillin (IM)**

- 1.2 million units > 7years
- 600 000 units < 7years of age
- Ask about family members with sore throat and treat.
- Educate family about B Pharyngitis

See section on safe administration of BPG

Why Benzathine Penicillin?

- Single injection
- Better bactericidal effect than oral
- Oral ttt needs **10 whole days** to be effective
- Oral macrolides: clinical improvement but no eradication of organism.
- Cost effective, evidence based.
- Parents and patients more satisfied.

Summary

- Simple clinical protocol for diagnosis of bacterial pharyngitis using one or 2 points

- Treat with BPG

Questions on Primary Prevention

1.The best way to diagnose Bacterial Pharyngitis (BPh) in patients 3-18 years of age in RHD endemic areas is by:

A. Clinical findings
B. Throat culture and rapid antigen test
C. High leukocyte count
D. High ESR

2. The best way to treat BPh in patients 3-18 years is:

A. Oral penicillin or cephalosporin for 5-7 days
B.IM Benzathine penicillin
C. Azithromycin for 3 days
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3. Primary prevention of acute rheumatic fever (ARF) means:

A. Prompt diagnosis and treatment of BPh
B. Giving 3 weekly penicillin
C. Diagnosis and treatment of ARF
D. Management of RHD

Case Study 1

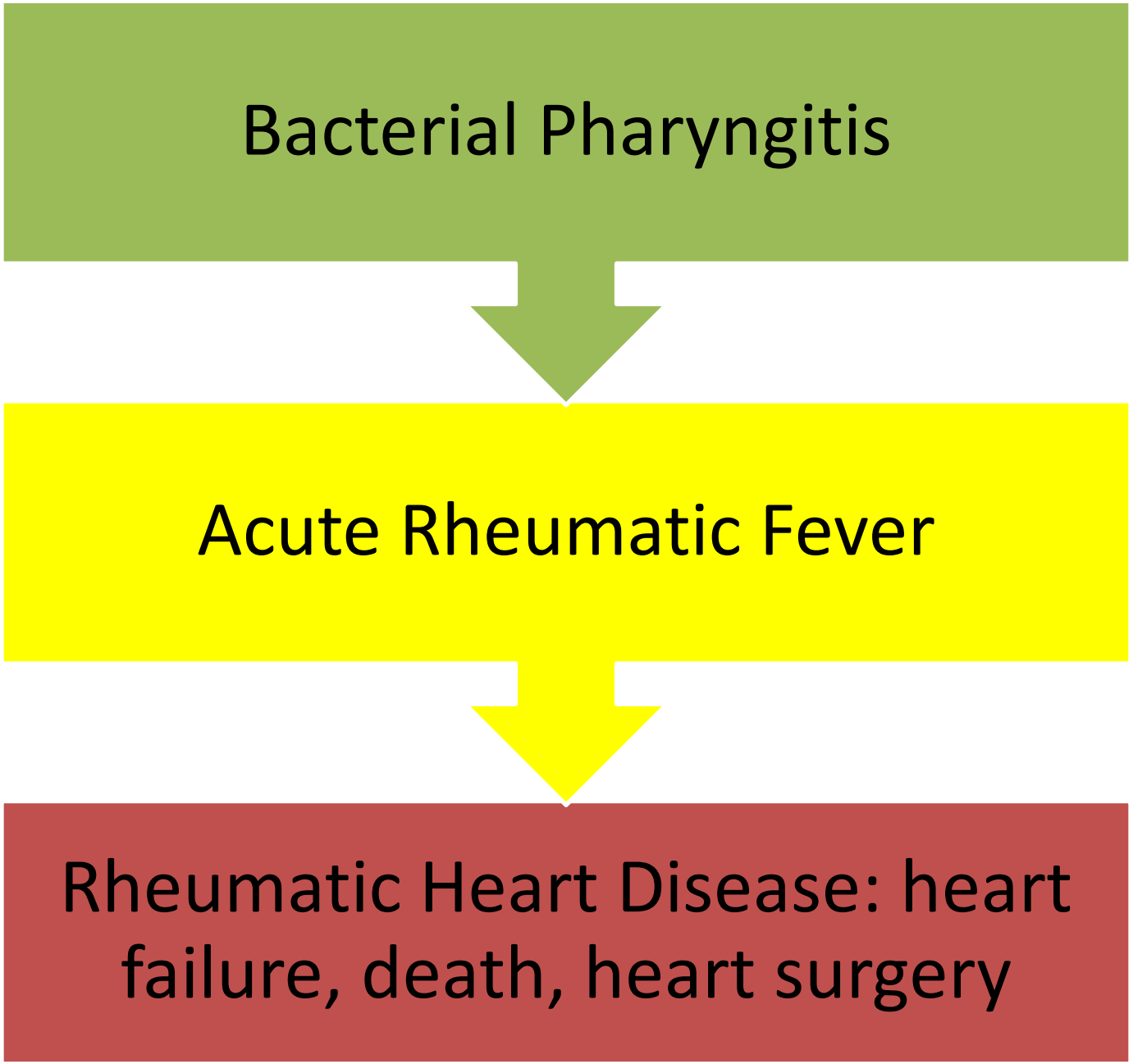
Adam is 7 years old complaining of sore throat for one day , which one of the following signs favors bacterial over viral pharyngitis:

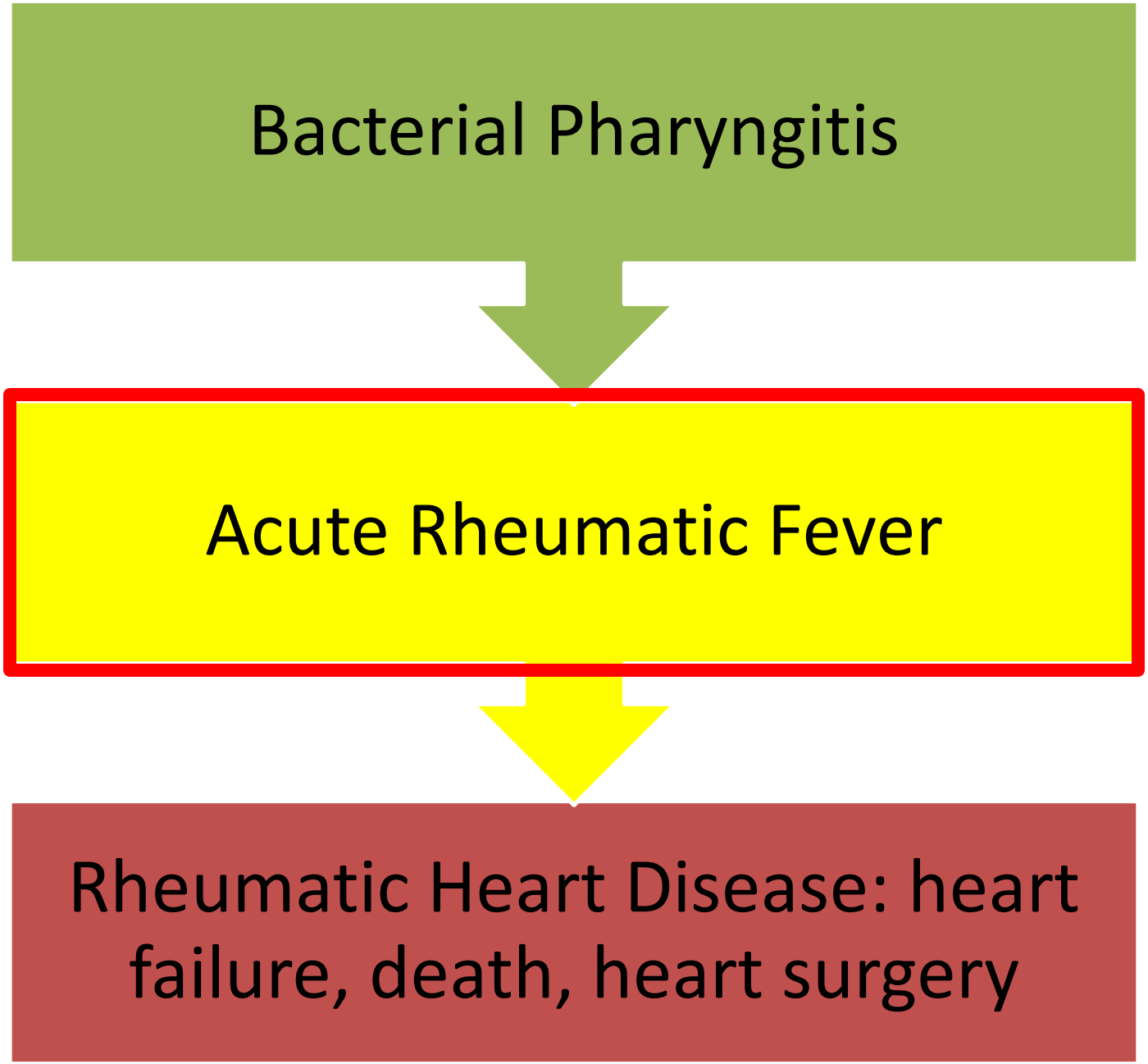
a. Horse voice
b. High ASO titre
c. Cough
d. Fever of 37.8 degrees
e. Absence of runny nose

Secondary Prevention

Definition

Early diagnosis and management of acute rheumatic fever (ARF)


Bacterial Pharyngitis
Acute Rheumatic Fever
Rheumatic Heart Disease: heart failure, death, heart surgery


Bacterial Pharyngitis
Acute Rheumatic Fever
Rheumatic Heart Disease: heart failure, death, heart surgery

Acute Rheumatic Fever (ARF)

- Complication of Strep Pharyngitis.
- Occurs in 0.3-2% of patients who have GAS pharyngitis

Diagnosis: The Jones Criteria

1. Major criteria
2. Minor criteria
3. Evidence of strep infection: ASO titre

Jones Criteria – 2015 Update

1. Inclusion of **subclinical carditis** (echo diagnosed) as a major criteria.
2. Monoarthritis as a major criteria
3. Polyarthralgia as a major criteria
4. Monoarthralgia as a minor criteria

Michael H Gewitz et al.Revision of the Jones Crtieria for diagnosis of acute rheumatic fever in the era of echo/Doppler.Circulation 2015;131:1806-18.

Jones Criteria (2015 Modification)

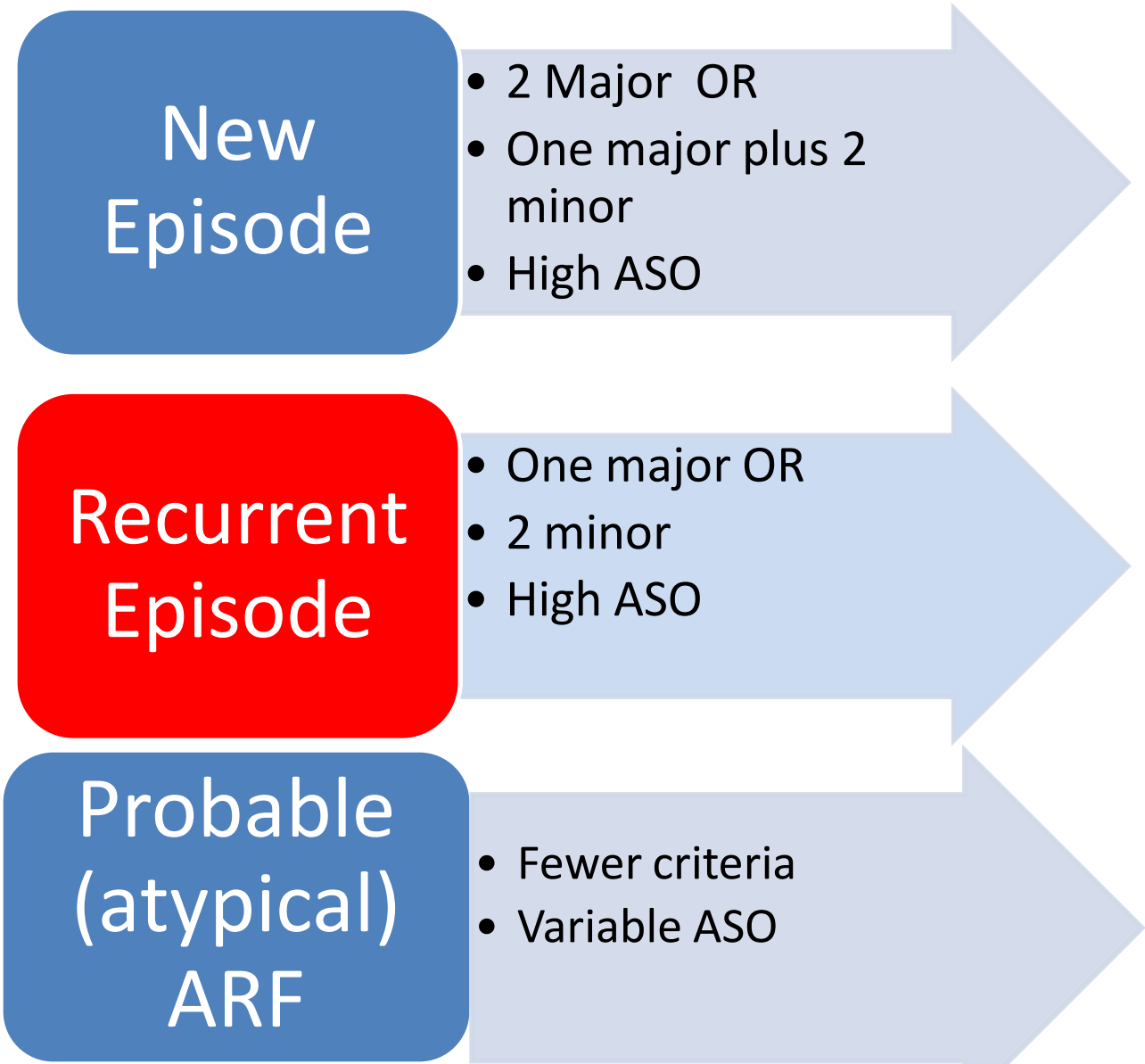
Major Criteria

1. Carditis (clinical or **echo diagnosed**)
2. Arthritis : polyarthritis ; **monoarthritis & polyarthralgia** in high risk areas
3. Chorea
4. Erythyma marginatum
5. Subcutaneous nodules

Minor Criteria

1. Fever
2. Polyrthralgia, **monoarthralgia** in high risk areas
3. Increased acute phase reactants
4. Prolonged PR interval

3 Categories of ARF



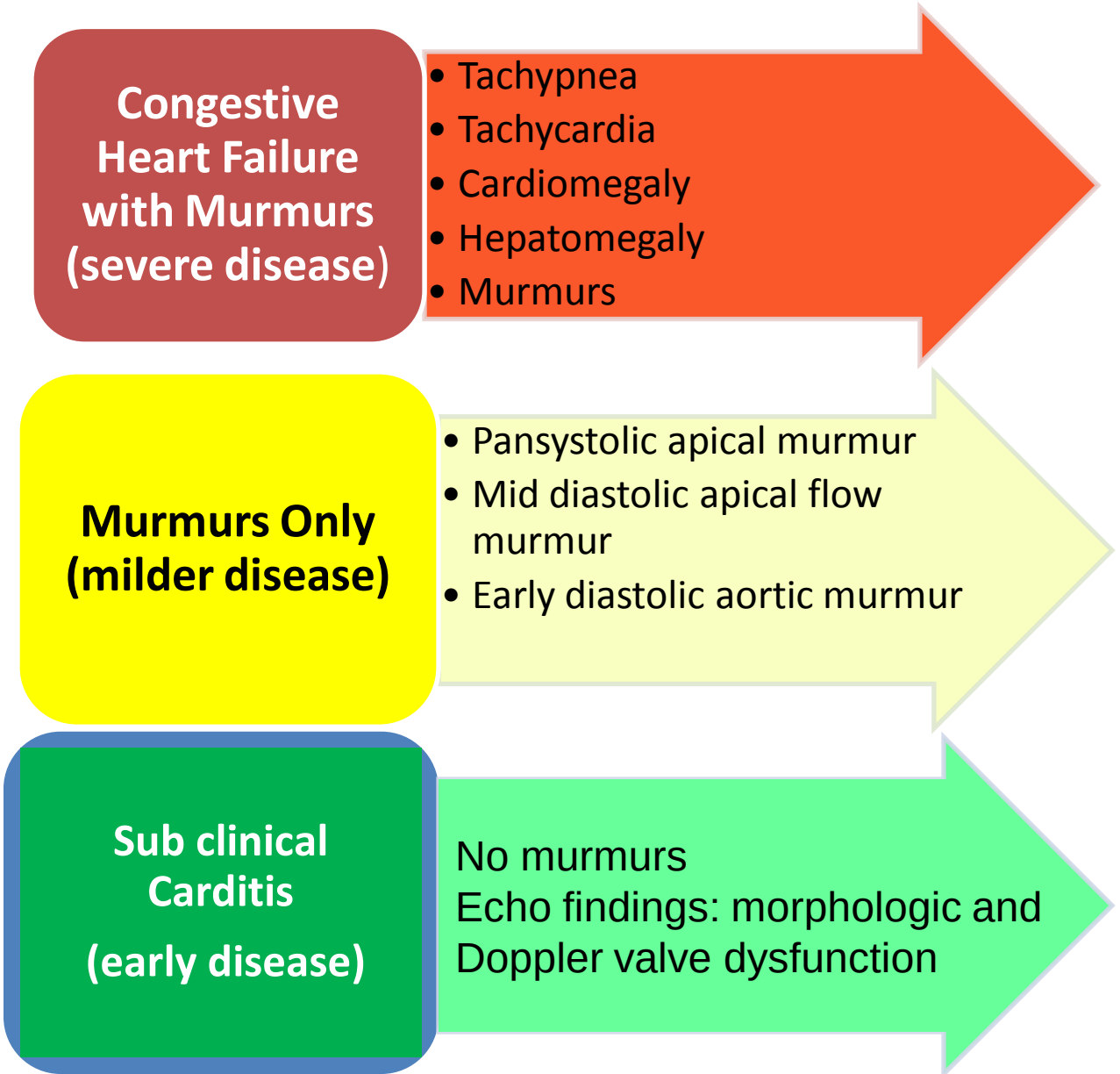
Major Criteria: Arthritis and polyarthralgia

- Migratory : Large joints
- Maximum severity in 12-24 hours, persists for 2-6 days
- Resolves spontaneously (dangerous as pt will not seek to medical care).

Major Criteria: Carditis

- Mitral regurgitation is the most common followed by combined aortic and mitral regurgitation.

Clinical Features of Carditis



Echo Features of Subclinical Carditis

Mitral

Morphologic Criteria

- Anterior Mitral valve leaflet thickening
- Chordae thickening
- Excessive leaflet tip motion
- Restricted Opening

Doppler Criteria

- MR jet => 2 cm
- Seen in 2 views
- Velocity ≥3 m/s for one complete envelope
- Pan systolic jet of MR

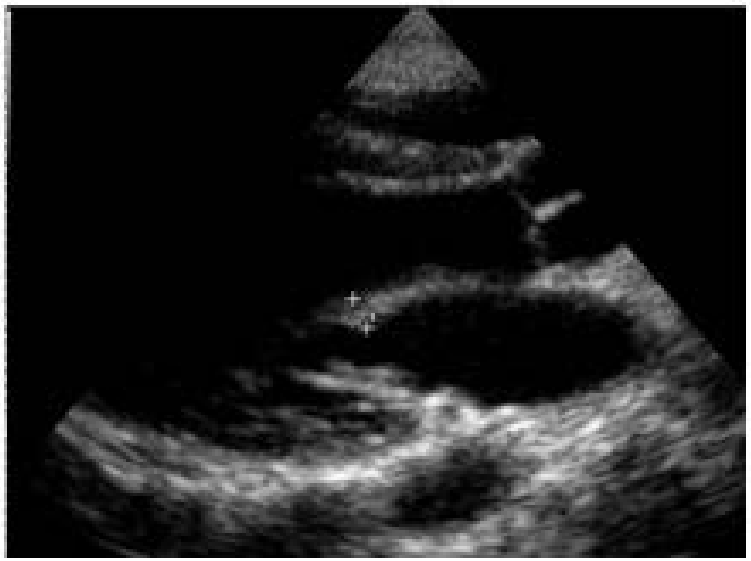
Echo Features of Subclinical Carditis Aortic

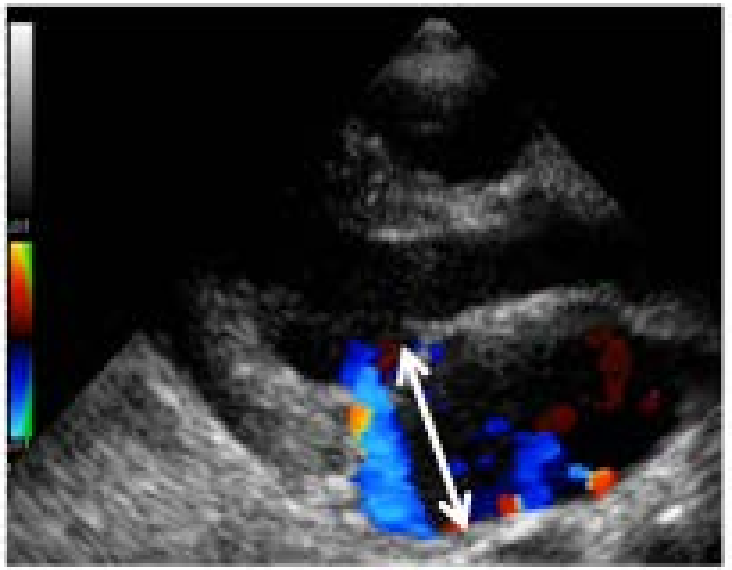
Morphologic Criteria

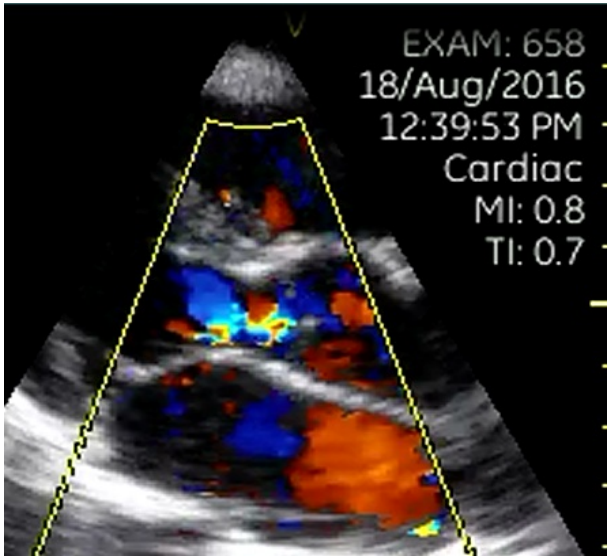
- Irregular thickening
- Cooptation defect
- Prolapse
- Restricted motion

Doppler Criteria

- AR jet => 1 cm
- Seen in 2 views
- Pan diastolic jet
- Velocity ≥3 m/s in early diastole






EXAM: 658
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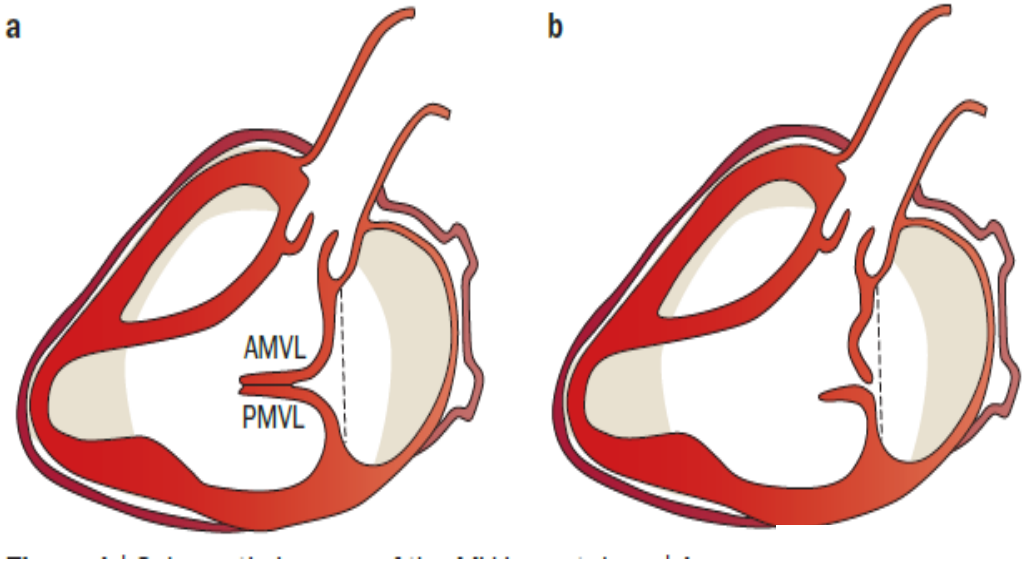
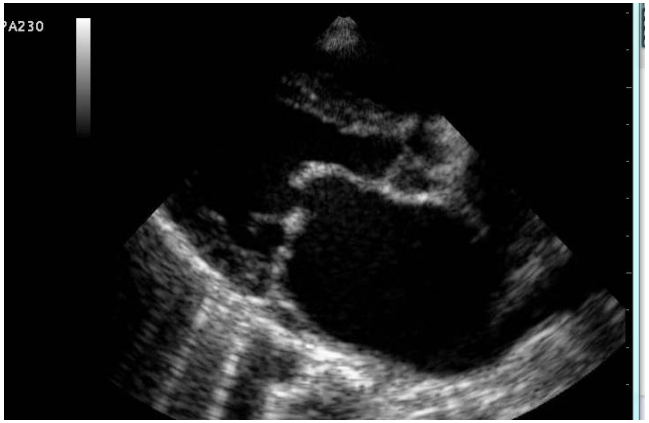
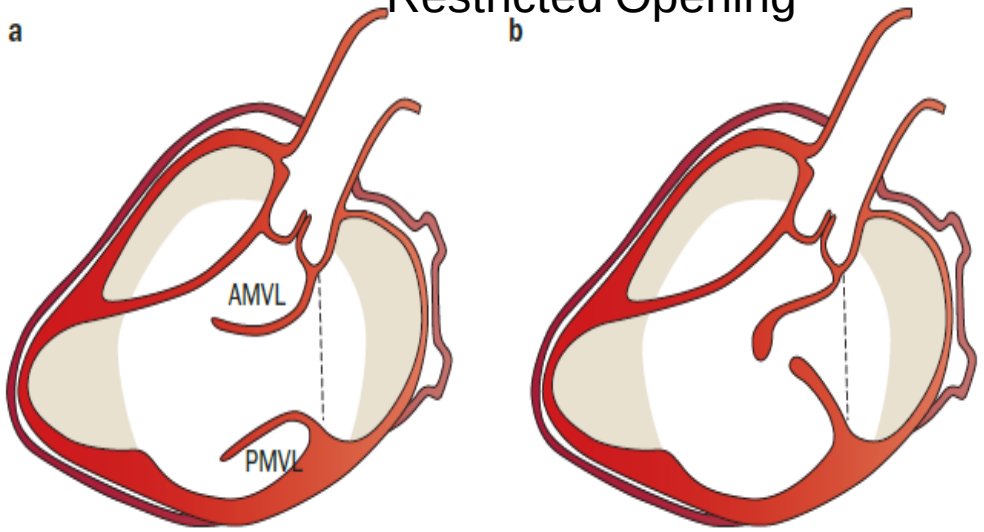
12:39:53 PM
Cardiac
MI: 0.8
TI: 0.7

Excessive leaflet tip motion



Restricted Opening





Reményi B, Wilson N, Steer A, Ferreira B, Kado J, Kumar K, Lawrenson J, Maguire G, Marijon E, Mirabel M, Mocumbi AO, Mota C, Paar J, Saxena A, Scheel J, Stirling J, Viali S, Balekundri VI, Wheaton G, Zühlke L, Carapetis J.World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease--an evidence-based guideline. Nat Rev Cardiol. 2012;9:297-309.

Diagnosis of Subclinical Carditis

Definite

-Doppler criteria plus 2 morphologic criteria

- MS mean gradient > 4 mmHg

-Borderline disease of both mitral and aortic valves

Borderline:

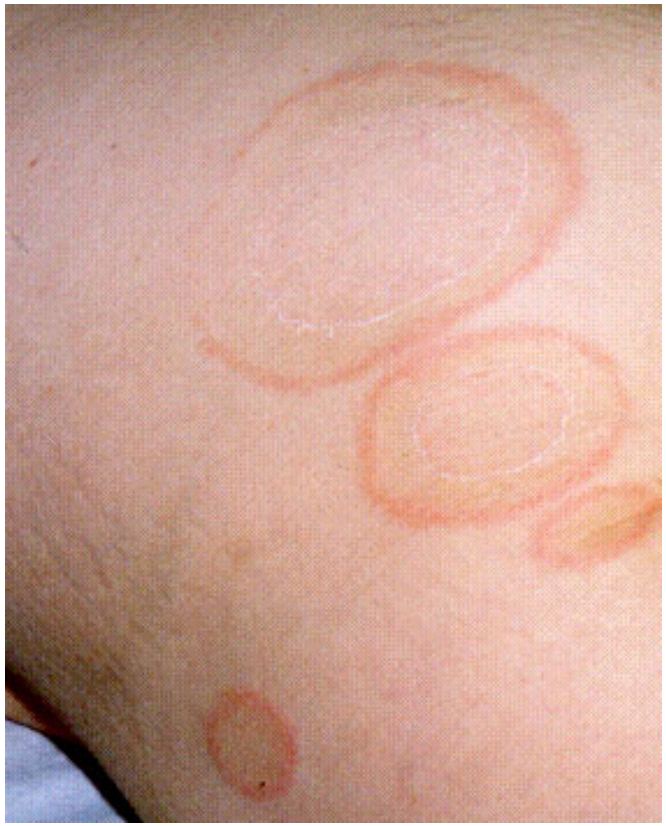
-Doppler criteria with no morphologic criteria

-2 Morphologic criteria

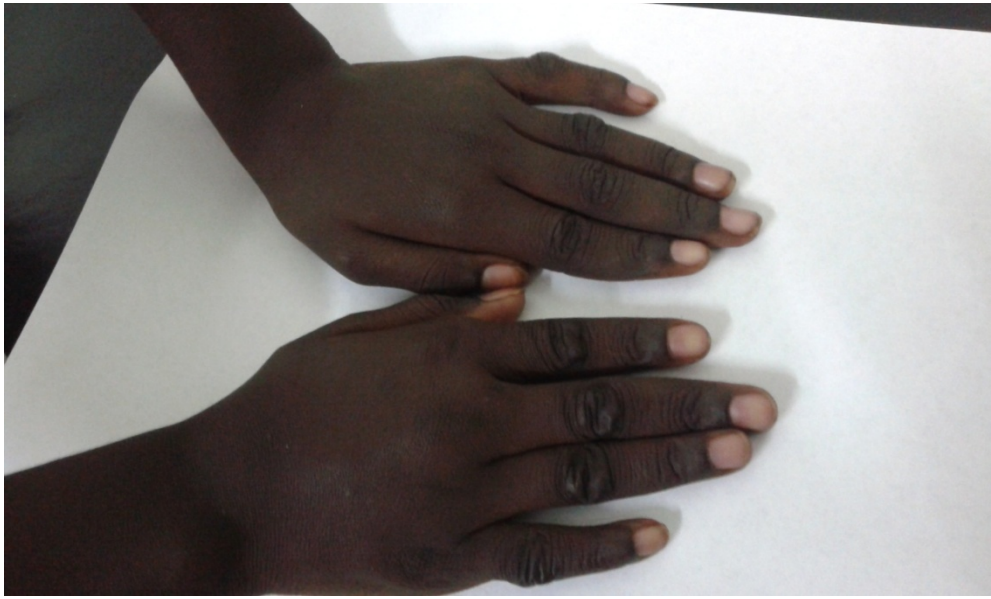
Sydenham's Chorea

- Involuntary, **purposeless** movements. emotional lability. Hyperextended joints, hypotonia, diminished reflexes
- *When present alone it is enough evidence of ARF.*

Erythema marginatum



Subcutaneous nodules



Management

Treat Infection: Benzathine penicillin stat and 3 weekly

Treat Inflammation :Aspirin in anti-inflammatory doses (75-100mg/kg/d) for 4-6 weeks.

- Steroids if aspirin is not tolerated or inflammatory markers not coming down

Supportive ttt: Bed rest, Treatment of heart failure.

Secondary Prevention

- Benzathine penicillin: **IM Q3 weeks**

  600 000 (<7y)

  1.2 million units (>7y)

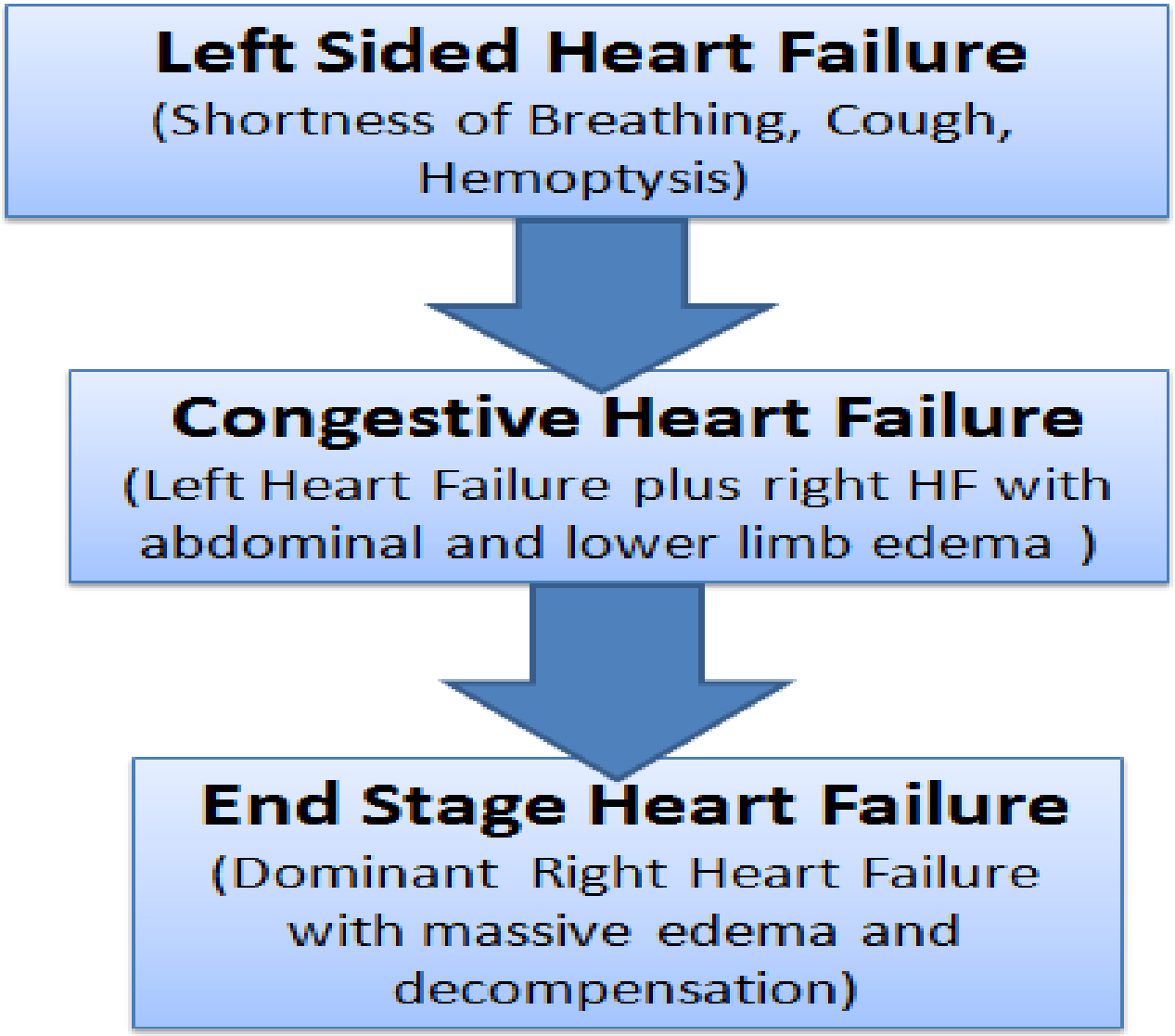
Duration of Prophylaxis

- Patients **without carditis**:

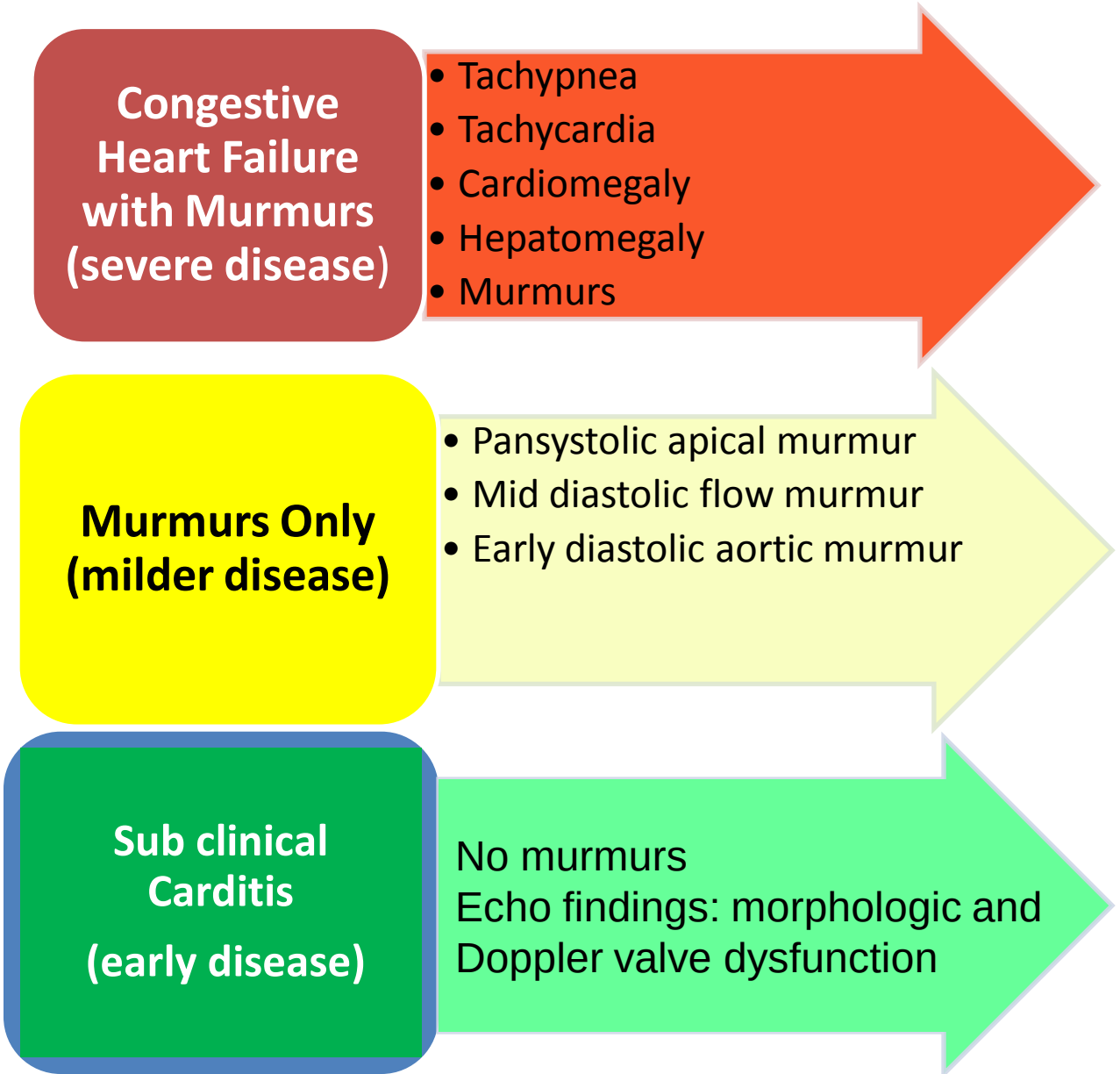
  up to 25 years of age
- Patients **with carditis**: and after valve surgery: Lifelong

Rheumatic Heart Disease

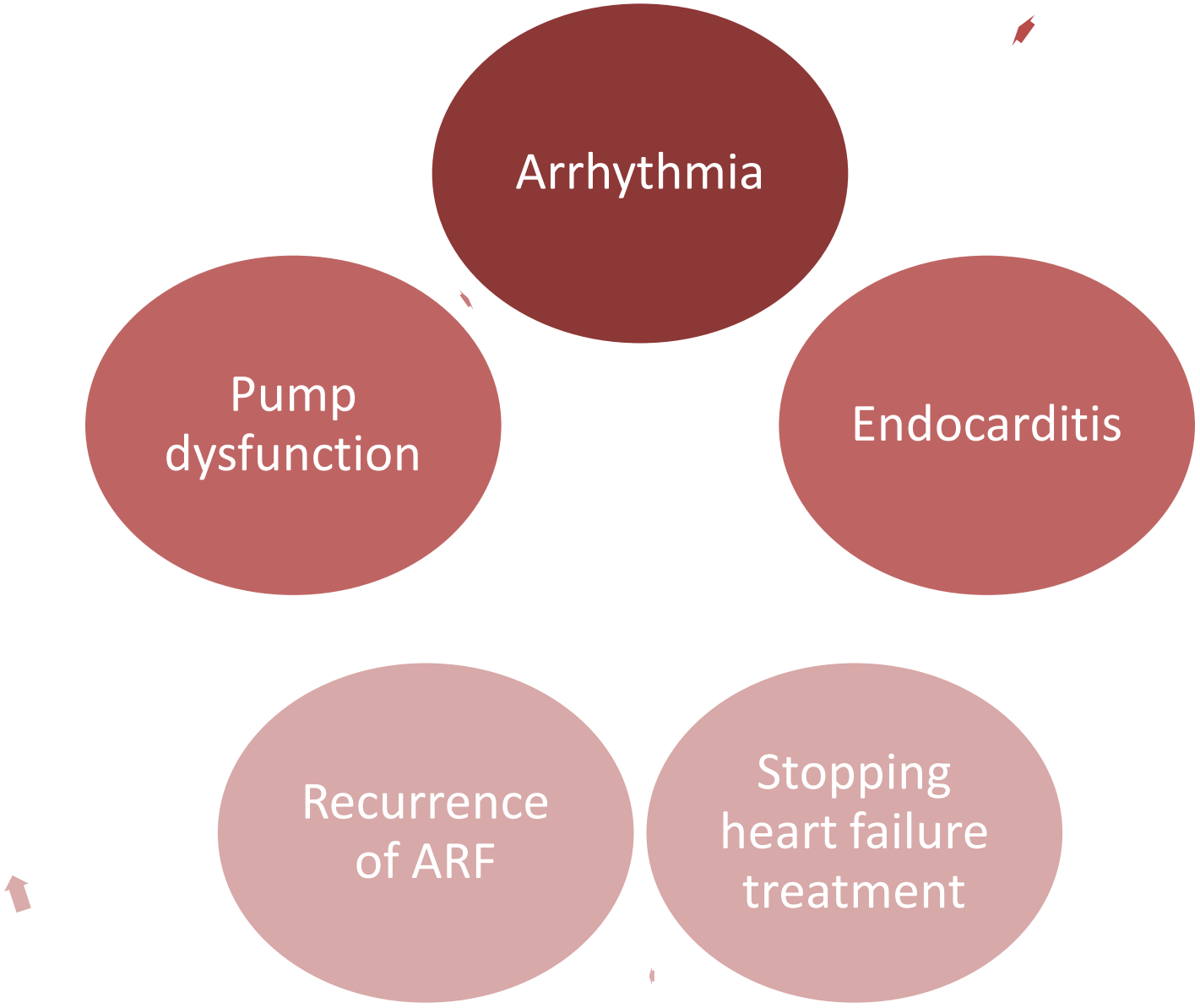
Patho physiology of RHD



Clinical Features of Carditis



Causes of Decomposition/Worsening



When you see a patient with RHD

R/O active disease (ARF):

Do ESR, CRP, ASO titre and consider ***recurrence*** of ARF if there is a major or 2 minor criteria and positive ASO

- **Recurrence of ARF is often missed**

Management of RHD

- **Medical :** anti heart failure treatment
- **Interventional:**

  - Transcatheter Balloon Dilatation: MS

  -Surgical: Valve repair

  Valve replacement

- Even after successful valve repair, recurrence of RHD can occur despite strict adherence to penicillin prophylaxis.

Post Valve Surgery

- Continuous follow up
- Don’t stop Benzathine Penicillin
- In patients with prosthetic valves:

Warfarin should not be stopped
INR control: target 2.5-3

- Endocarditis prophylaxis
- Dental hygiene

Case Study (2)

Sara is 15 year old girl who has been diagnosed as RHD 2 years ago, she presented with ankle pain for 2 days, which of the following is true:

a. If there is leucocytosis and high ASO, she should receive aspirin in a high dose
b. Recurrence of ARF needs to be considered only if she has fever.
c. We need 2 major criteria to diagnose recurrence of ARF
d. If she is compliant with BPG , no need to request further investigations
e. Ankle pain is considered a minor Jones criteria

Safe Administration of Benzathine Penicillin

Problems of BPG can be solved

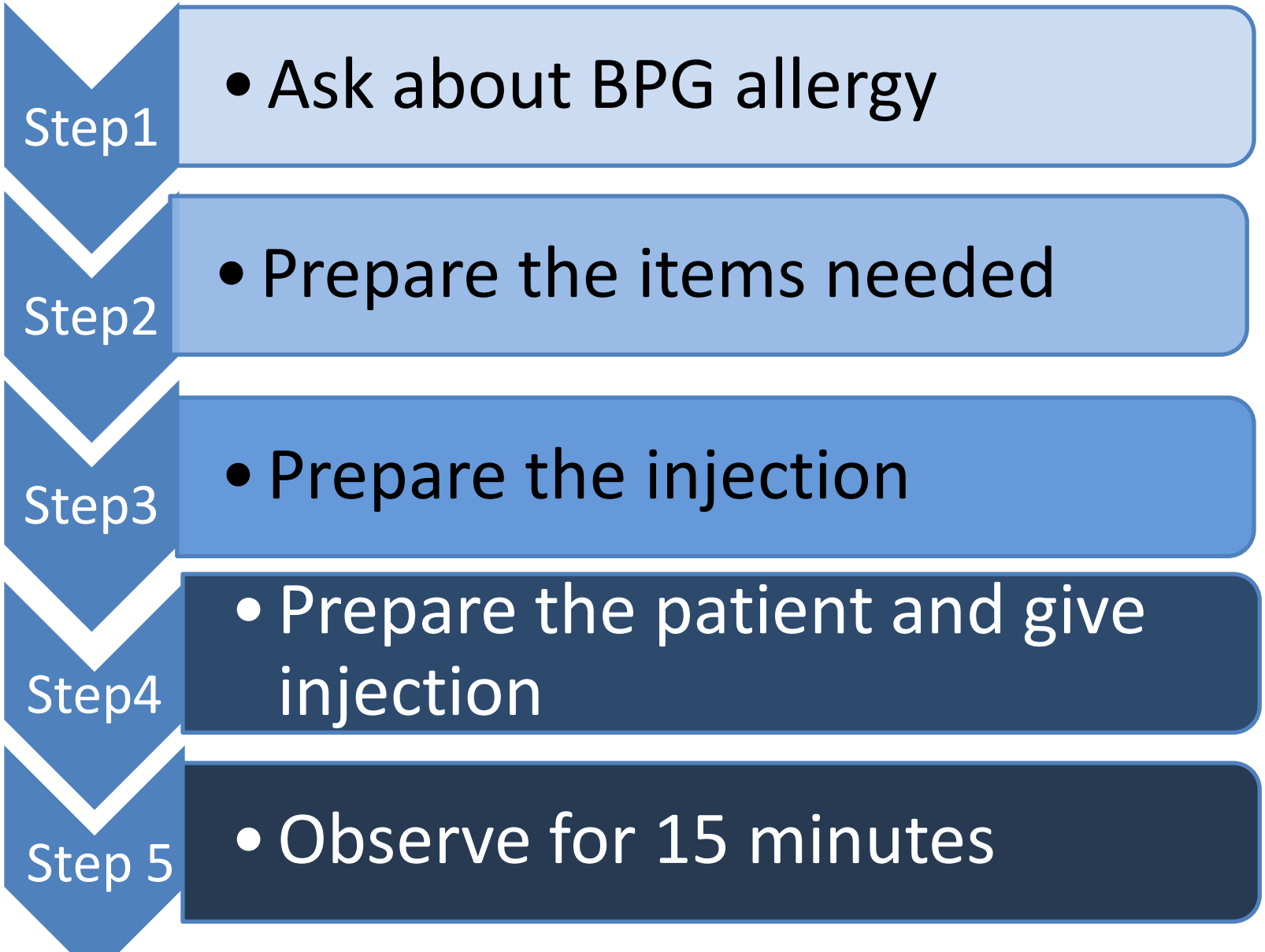
Problem	Solution
The drug is "heavy"	Use appropriate amount of diluents at room temp
The drug can block the needle	Use a large bore needle
The drug is painful	Dilute the powder in lidocaine 2% and inject slowly
Patients fear allergy	Serious allergy is very rare-reassure patients
Health workers fear allergy	Training of health workers on allergy management help them to be confident

Important

- Skin testing using diluted BPG has **NO ROLE** in prediction of allergic patients
- Don’t perform this Skin testing
- Go by the 5 Steps Protocol

Aiden Long (Associate Professor of Immunology, Harvard Medical School, USA) .Do we need to do skin testing using dilute BPG?. PASCAR RHD Committee Meeting, Cairo, 2017

5 Steps for BPG Administration



Step 1:Ask about the H/O Allergy

H/O Severe allergy (anaphylaxis)

- DON'T GIVE BPG
- Give Erythromycin

H/O Mild Allergy (hives, itching)

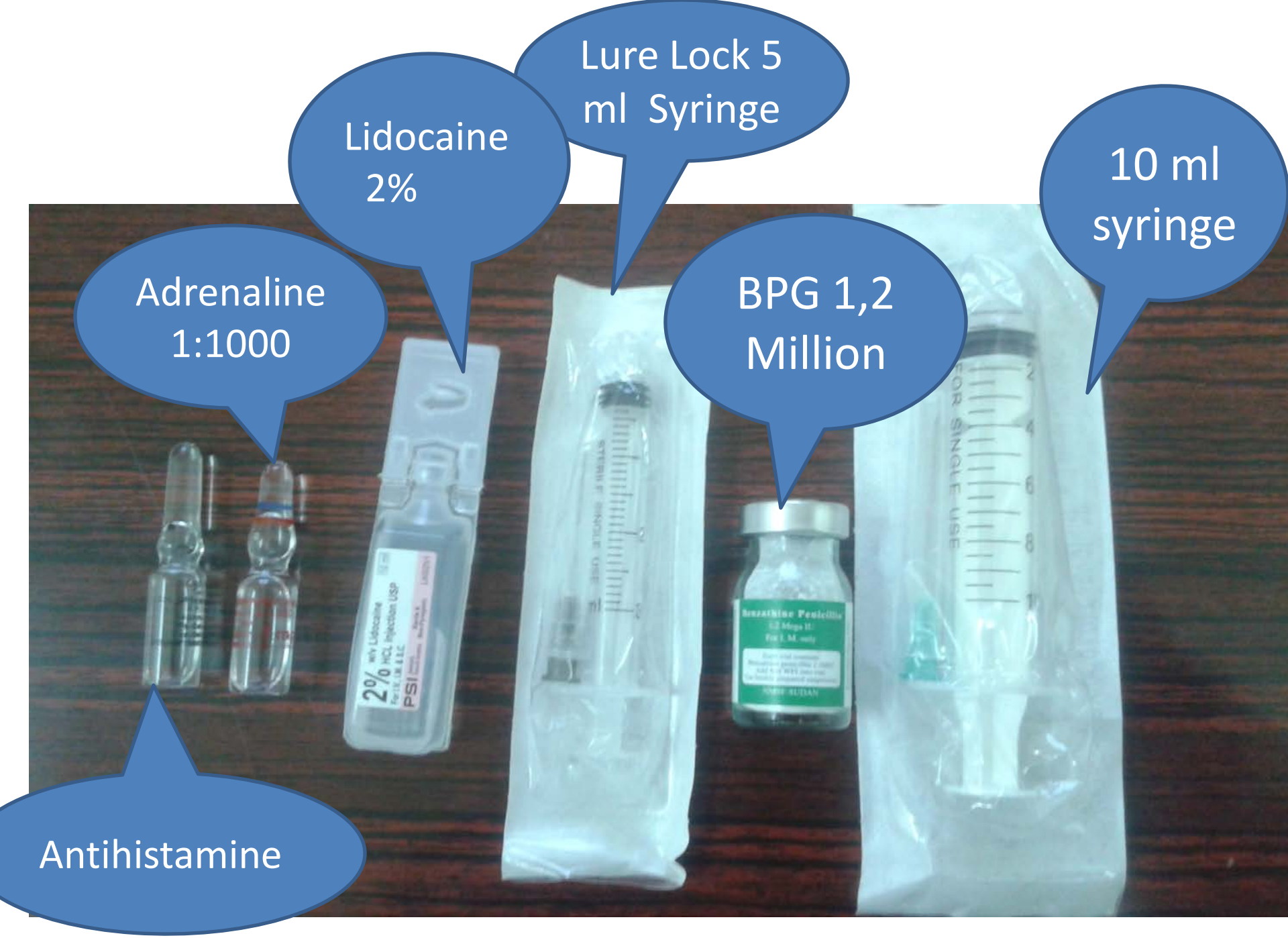
- **Give a TEST DOSE: 1/10th** of the dose IM
- **Observe:** If no reaction: Give the rest of the dose
- If there is reaction: Treat the reaction and give Erythromycin

No H/O allergy

Give BPG

Step 2: Prepare the following

1. One 10 ml syringe
2. One 5 ml syringe (Lure Lock)
3. One BPG ampoule 1.2 million units
4. One vial of local anesthetic lidocaine (Lignocaine) 2% (or water for injection)
5. One adrenaline vial 1:1000
6. One antihistamine vial


Lure Lock 5 ml Syringe
Lidocaine 2%
10 ml syringe
Adrenaline 1:1000
BPG 1,2 Million
Antihistamine

Step 3: Prepare the injection:

- Draw appropriate amount of local anesthetic as diluents for the BPG powder (make sure it's not cold)
- Inject the diluent into the BPG vial

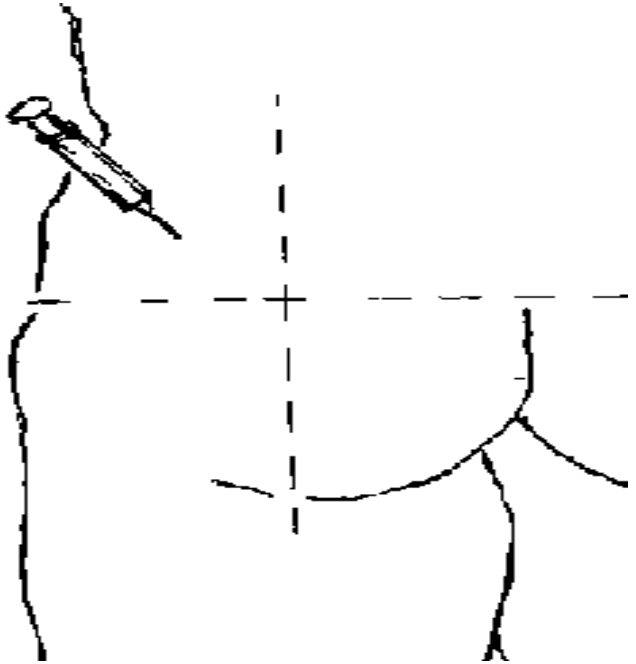
Mix gently till dissolved

Draw in a 5 ml syringe

Insert the large bore needle of the 10ml syringe

Step 4: Prepare the patient and give the injection:

- Ask the patient to lie on the abdomen
- Mark the site of the injection on the gluteus muscle (Figure)
- To minimize pain: press with your thumb over the site for 10 seconds
- Aspirate first to avoid veins then give slowly



Dose:

- For patients 7 years of age or more: 1.2 million units
- For patients less than 7 years of age: 600 000 units

IMPORTANT

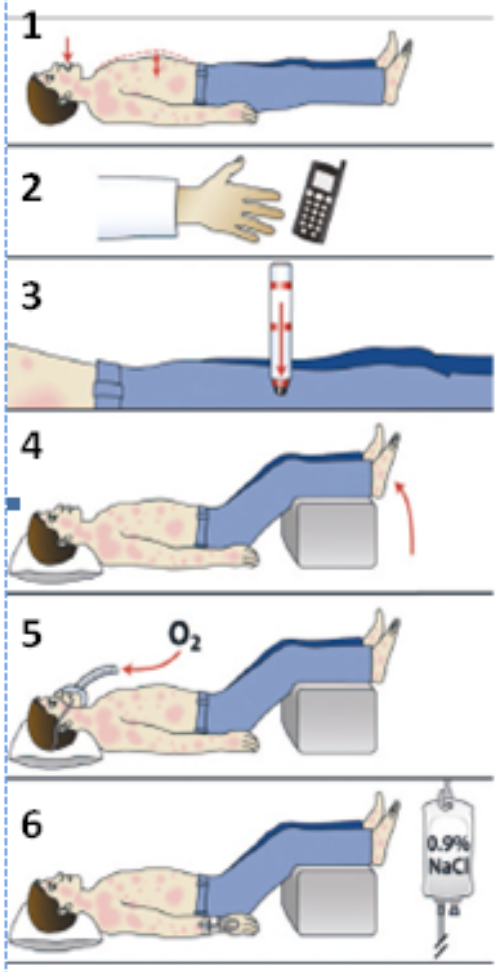
NEVER EVER GIVE INTRAVENOUS THIS LEADS TO IMMEDIATE MORTALITY

Step 5: Observe and treat reaction

- Observe for 15 minutes
- If an allergic reaction develops:

- Local Reaction :Itching, hives:
    - Antihistamine injection
    - Continue observation till well

Systemic : Anaphylaxis



1.Assess ABC: if needed perform CPR

DO STEPS 2,3,4 QUICKLY AT SAME TIME

2.Call for help

3.Inject adrenaline: 0.3 ml (<7 y), 0.5 ml (>7 y) of 1:1000 solution. Can be repeated in 15 minutes

4.Lie the patient with legs up

5.If distressed, give O2

6.IV line: give IV normal saline10ml /kg boluses. Can give adrenaline infusion

World Allergy Organization Guidelines for anaphylaxis management. WAO2011;4:13-37

For Penicillin sensitive Patients

- **Erythromycin** BD for 10 days (for treatment) and for the duration of secondary prophylaxis

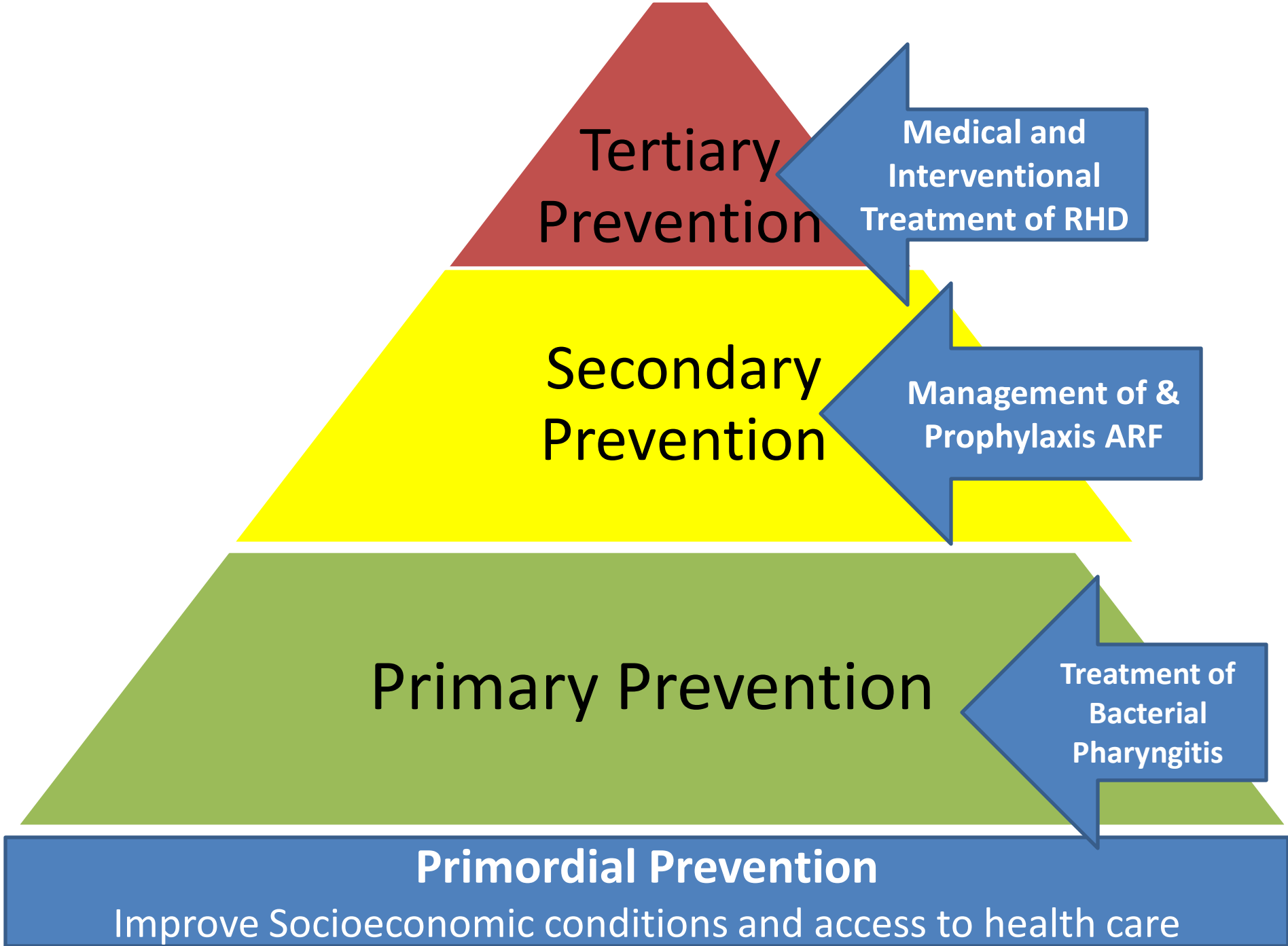
Dose:

- Less than 7 years: 250 mg BD for 10 days (for treatment) and for the duration of secondary prophylaxis
- More than 7 years: 500 mg BD for for 10 days (for treatment) and for the duration of secondary prophylaxis.

RHD in Pregnancy

- Important cause of maternal mortality
- All girls with RHD should be counseled before pregnancy
- Mild lesion: no problem, F/U and regular use of penicillin
- Severe lesion: contraception, treat lesion before pregnancy
- Early referral for tertiary center

RHD Control Program

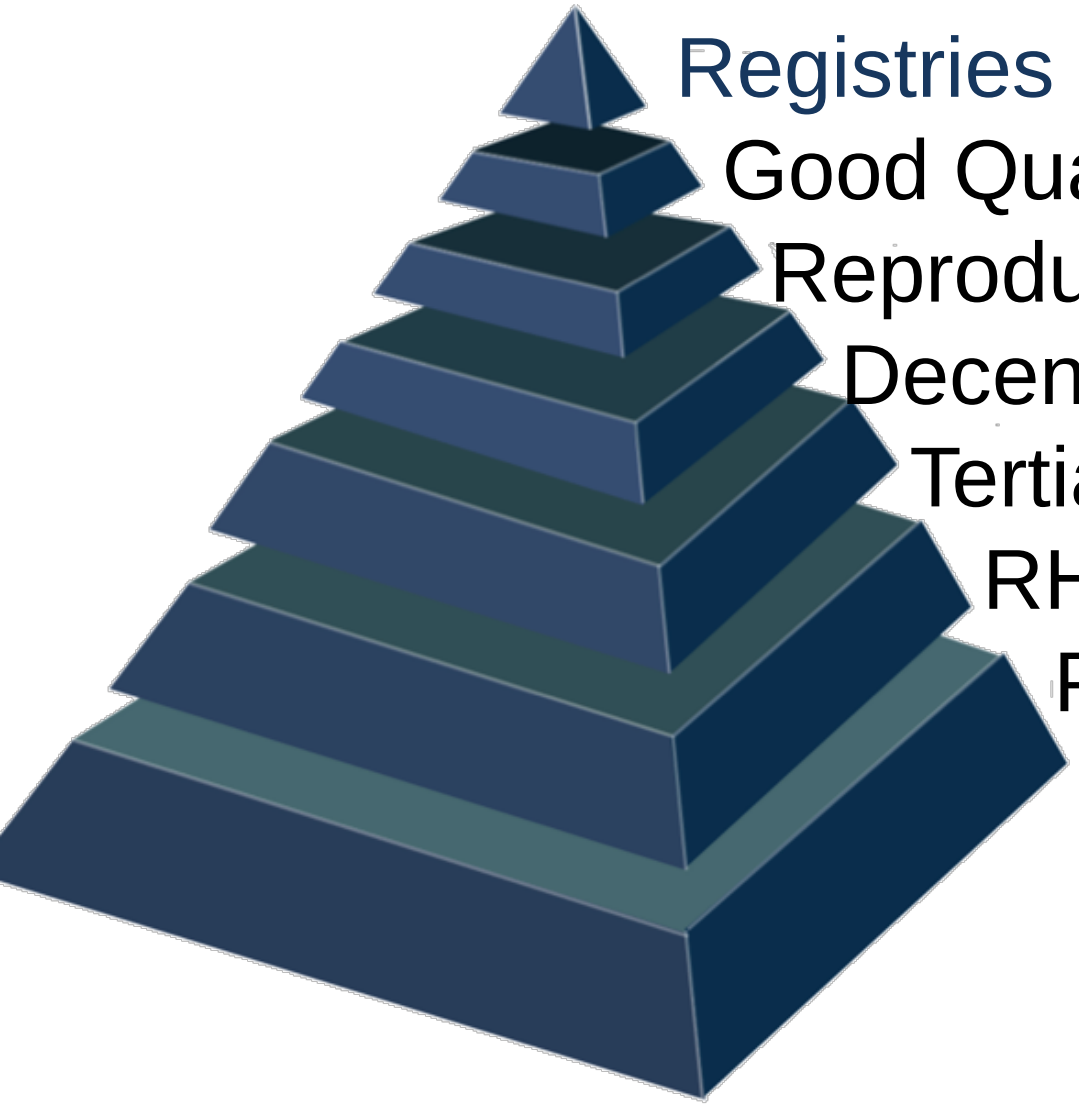

Tertiary Prevention
Medical and Interventional Treatment of RHD
Secondary Prevention
Management of & Prophylaxis ARF
Primary Prevention
Treatment of Bacterial Pharyngitis
Primordial Prevention
Improve Socioeconomic conditions and access to health care

Objectives

- To reduce RHD in those <25 years of age by 25% by the year 2025.

Bo Remenyi, Jonathan Carapetis, Rosemary Wyber, Kathryn Taubert and Bongani M. Mayosi.Position statement of the World Heart Federation on the prevention and control of rheumatic heart disease. Nat. Rev. Cardiol2013; 10:284–292.

The 7 Key Actions to eradicate RHD



David Watkins, Liesl Zuhlke, Mark Engel et aSeven key actions to eradicate rheumatic heart disease in Africa: the Addis Ababa communiquél.Cardiovas J Africa 2016

Steps to Control RHD

(1)

Form a committee with all stakeholders

(1)

Form a committee with all stakeholders

(2)

Training Material (Guidelines and Modules)

(1)

Form a committee with all stakeholders

(2)

Develpe Training Material (Guidelines and Modules)

(3)

Integrate into MOH programs

(1)

Form a committee with all stakeholders

(2)

Develp Training Material (Guidelines and Modules)

(3)

Integrate into MOH programs

(4)

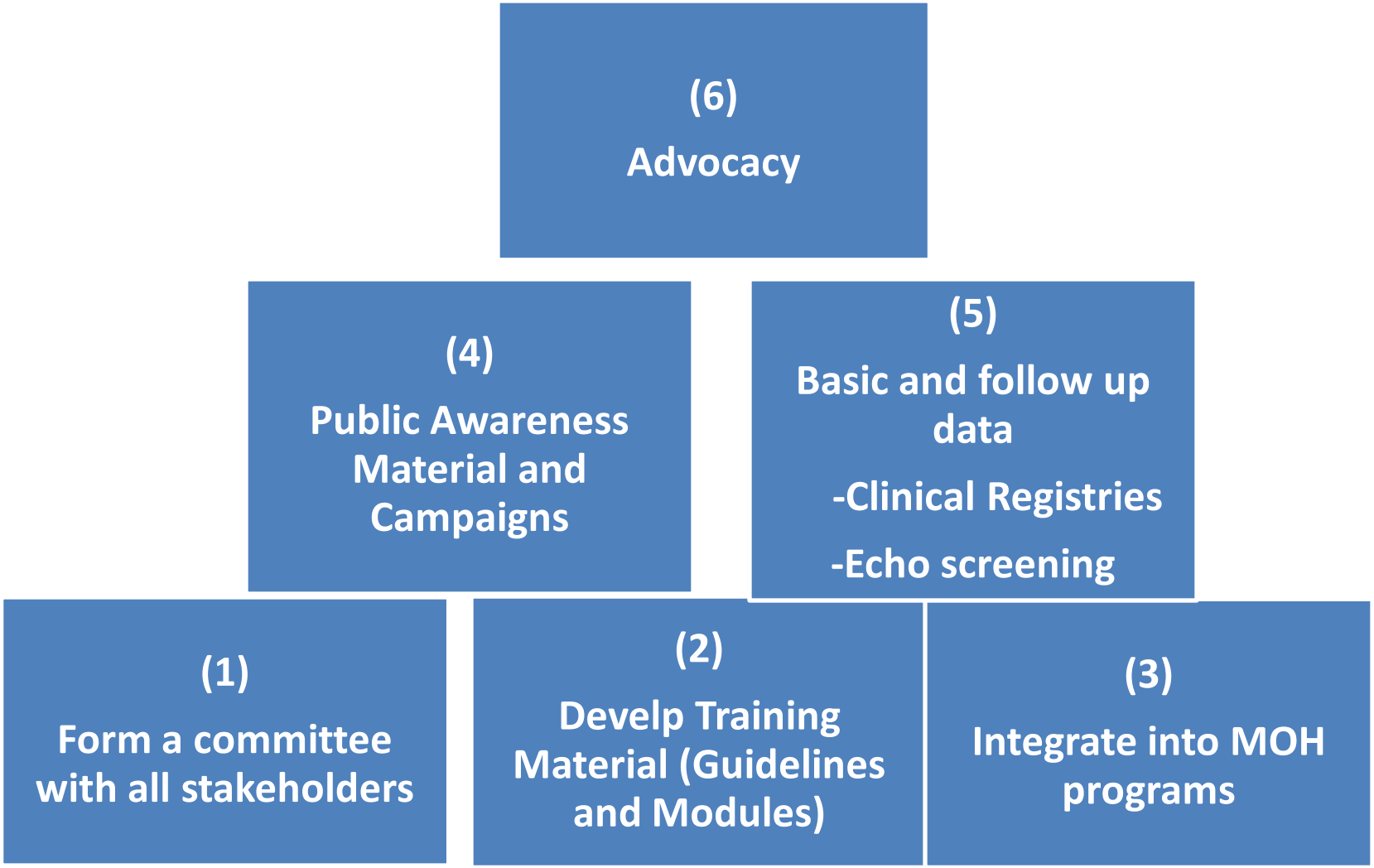
Public Awareness Material and Campaigns

(5)

Basic and follow up data

-Clinical Registries

-Echo screening


(6)
Advocacy
(4)
Public Awareness Material and Campaigns
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Basic and follow up data
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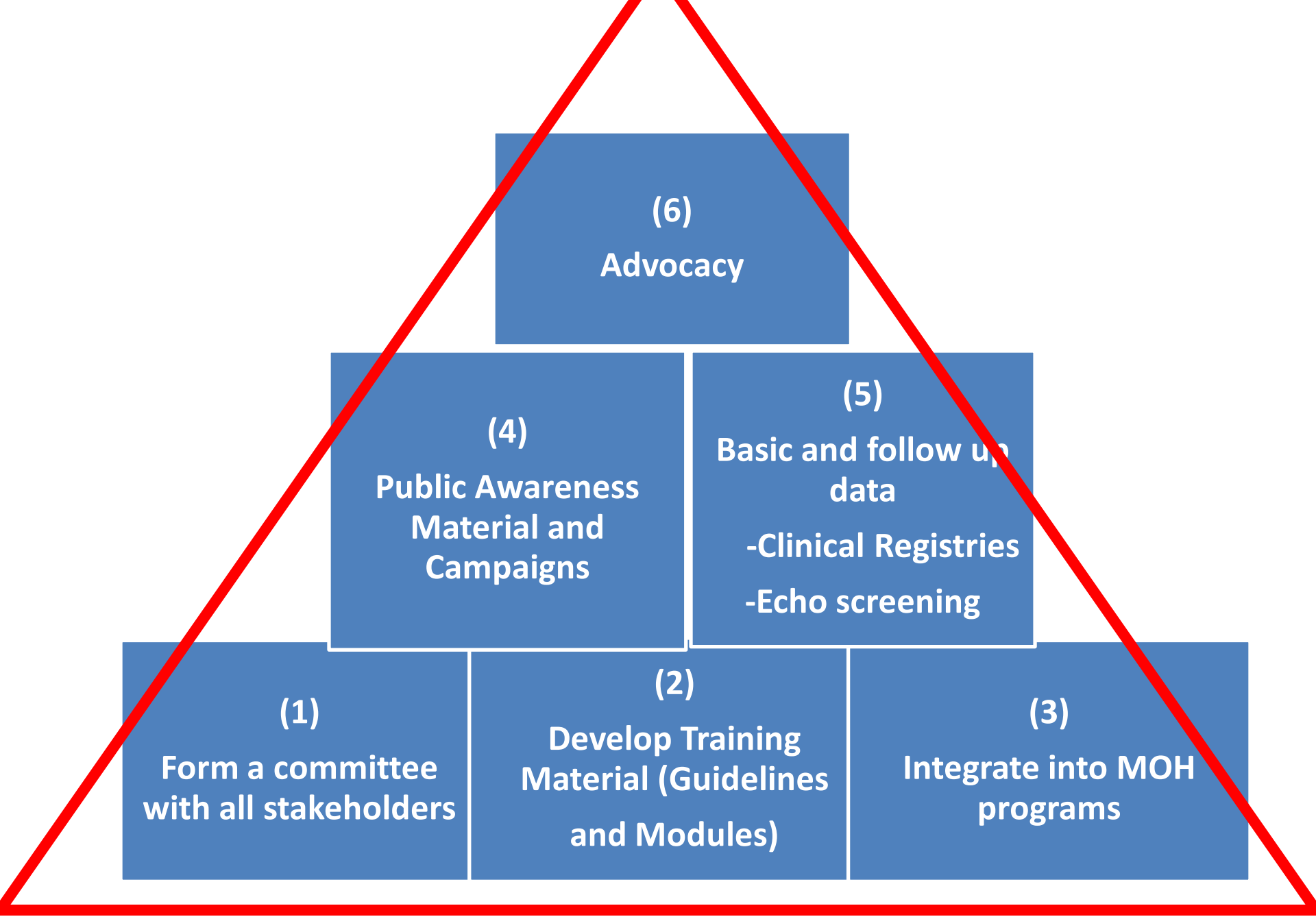
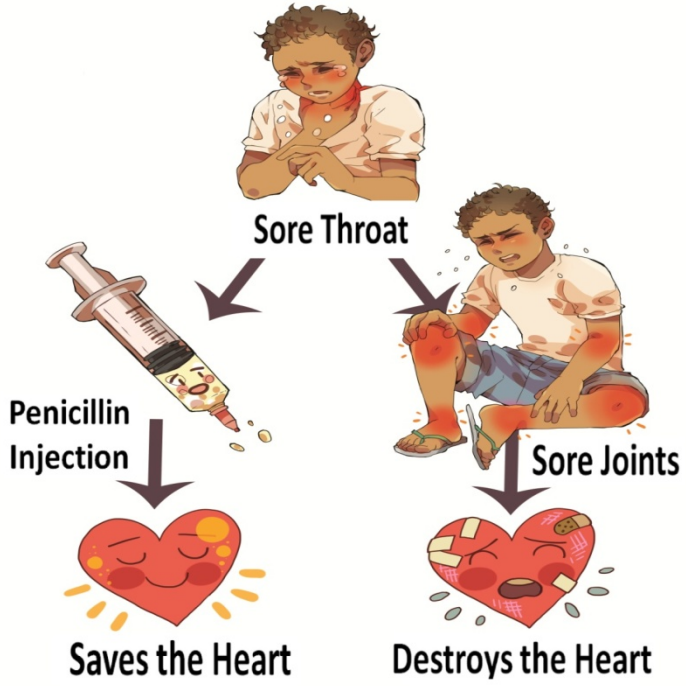
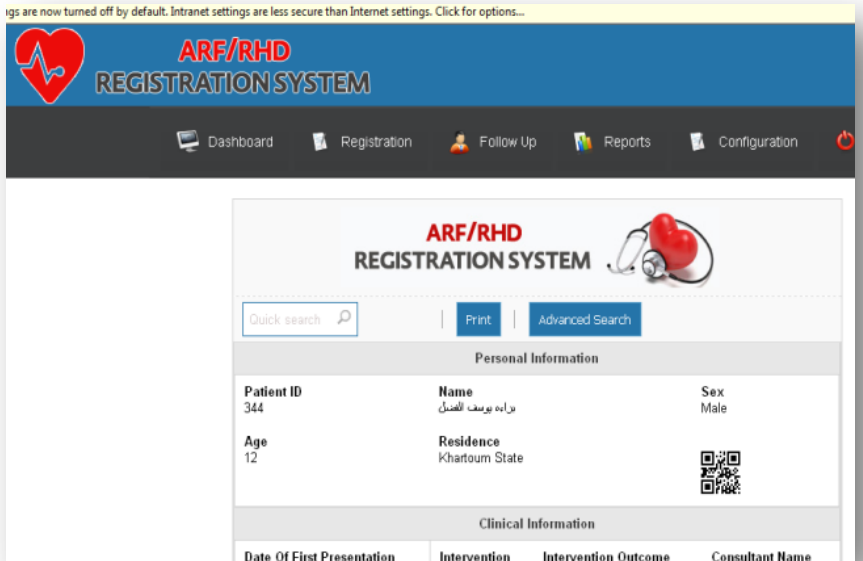

(6)
Advocacy
(4)
Public Awareness Material and Campaigns
(5)
Basic and follow up data
-Clinical Registries
-Echo screening
(1)
Form a committee with all stakeholders
(2)
Develop Training Material (Guidelines and Modules)
(3)
Integrate into MOH programs

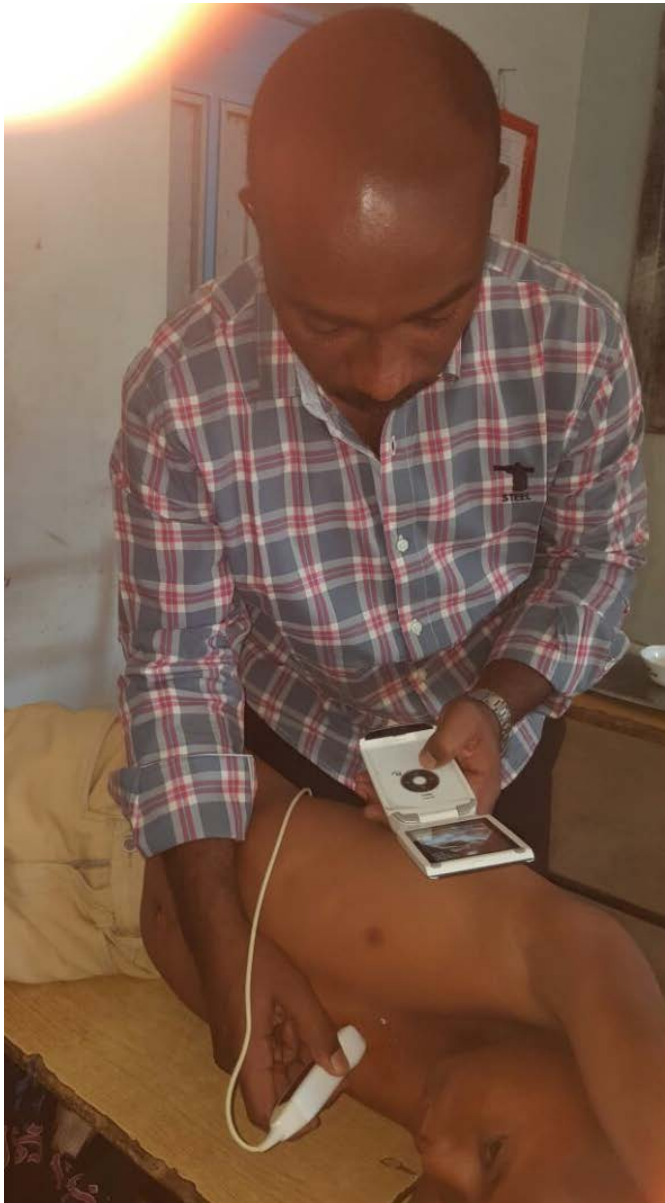
Public Awareness



Clinical and Echo Data

- Early detection
- Echo screening
- Registries: Manual or electronic





Health Workers’ Training

Summary

1. Jones criteria were modified to include monoarthritis, polyarthralgia and echo as major and monoarthralgia as minor criteria.
2. New entity of Borderline ARF is introduced
3. management of ARF: High dose aspirin for 4 weeks, & BPG q 21 days
4. Use the **5 Step Guidelines** for BPG administration, no need for skin testing.

Writing Committee

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Last updated: May 22/ 2017