

# Rheumatic Fever & Rheumatic Heart Disease

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# Lecture Outline

- ❑ What is ARF & RHD?
- ❑ Diagnosis
- ❑ Jones Criteria
- ❑ Differential Diagnosis
- ❑ Investigations, Management
- ❑ Rheumatic Valvular Heart Disease
- ❑ Prevention

# Acute Rheumatic Fever (ARF)

- ❑ Follows group (A)  $\beta$ -hemolytic streptococcal throat infection
- ❑ Represents a delayed immune response to infection with manifestations appearing after a period of 2-4 weeks
- ❑ Age 5-15 yrs
- ❑ A multisystem disease
- ❑ RHD is a long term complication of ARF
- ❑ Major effect on health is due to damage to heart valves

# Global Burden of RHD-WHO

- ❑ A leading cause of CV morbidity & mortality in young people
- ❑ Total cases with RHD 15.6 Millions
- ❑ Annual incidence of RF: 0.5 Million, nearly half develop carditis
- ❑ Estimated deaths from RHD: 250,000/YR
- ❑ Imposes a substantial burden on health care systems with limited budgets

# Epidemiologic Background

- ❑ The incidence of RF and the prevalence of RHD has declined substantially in Europe, North America and other developed nations
- ❑ This decline has been attributed to improved hygiene, reduced household crowding, and improved medical care

# Epidemiologic Background

- ❑ The major burden is currently found in low and middle income countries (India, middle east, gulf countries), and in selected indigenous populations of certain developed countries (Australia and New Zealand).
- ❑ A disease of poverty and low socioeconomic status
- ❑ In underdeveloped countries RHD is the leading cause of CV death during the first five decades of life

# ARF & RHD in Saudi Arabia

- ❑ Published data in KSA are limited.
- ❑ In developed countries the incidence of ARF has declined over past 50 yrs, incidence ranging 0.2 -0.64/100,000 (USA).
- ❑ ARF incidence in Eastern province was 22/100,000, age 5-14 yrs.
- ❑ A large study from Western province in 1991, showed a prevalence of RHD 2.4/1000, and an overall prevalence of RF and RHD 3.1/1000, age 6-15 yrs.





# Diagnosis of ARF

- ❑ No single test to diagnose ARF
- ❑ The symptoms and signs are shared by many inflammatory and infectious diseases
- ❑ Accurate diagnosis is important
- ❑ **Overdiagnosis** will result in individuals receiving treatment unnecessarily
- ❑ **Underdiagnosis** may lead to further episodes of ARF causing damage, and the need for valve surgery, CHF and death

# Diagnosis of ARF

- ❑ Diagnosis is primarily clinical and is based on a constellation of signs and symptoms and lab findings, which were initially established as the Jones criteria
- ❑ In 1944 Dr. TD Jones published a set of guidelines for diagnosis of ARF “Jones Criteria”
- ❑ Subsequently revised in 1965, 1984, 1992 and recently on 2015 by AHA

# Carditis

- ❑ Occurs in 50-70% of cases
- ❑ Only manifestation of ARF that leaves permanent damage
- ❑ May be subclinical
- ❑ Murmurs of MR or AR may occur in acute stage while mitral stenosis occurs in late stages
- ❑ Cardiomegaly and CHF may occur

# Arthritis

- ❑ Common: present in 35-66%
- ❑ Earliest manifestation of ARF
- ❑ Large joints: The knees and ankles, shoulders, elbows
- ❑ “Migrating”, “Fleeting” polyarthrititis
- ❑ Duration short < 1 week
- ❑ Rapid improvement with salicylates
- ❑ Does not progress to chronic disease

# Sydenham Chorea

- ❑ Also known as Saint Vitus'dance
- ❑ Occur in 10-30%, extrapyramidal manifestation, female predominance
- ❑ Abrupt Purposeless involuntary movements of muscles of face, neck, trunk, and limbs.
- ❑ Delayed manifestation of ARF -months
- ❑ Clinically manifest as-clumsiness, deterioration of handwriting,emotional lability or grimacing of face

# Subcutaneous Nodules

- ❑ Occur in 10%
- ❑ Usually 0.5 – 2 cm long
- ❑ Firm non-tender
- ❑ Occur over extensor surfaces of joints, on bony prominences, tendons, spine
- ❑ Short lived: last for few days
- ❑ Associated with severe carditis



# Erythema Marginatum

- ❑ Occurs in < 5%
- ❑ Highly specific for ARF
- ❑ Reddish border, pale center, round or irregular serpiginous borders, non-pruritic, transient rash
- ❑ Occurs on trunk, abdomen or proximal limbs
- ❑ Associated with carditis



# Diagnosis

- ❑ The Jones Criteria for diagnosis of ARF were first published by T Duckett Jones in 1944.
- ❑ Criteria underwent major revisions by AHA in 1992 and in 2015



# Revised Jones Criteria

## 1992

Major criteria	Minor criteria
Carditis	Hyperpyrexia
Arthritis	Arthralgia, without other signs of inflammation
Chorea	Laboratory indicators of acute phase:
Erythema marginatum	ESR, CRP
Subcutaneous nodules	Prolonged PR interval in ECG

And evidence of antecedent streptococcal infection

## 2015

### **Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography**

**A Scientific Statement From the American Heart Association**

*Circulation.* published online April 23, 2015;

# 2015 Revision of Jones Criteria

1) In accordance with the degree of **prevalence** of ARF/RHD in the population:

- ❑ **Low risk populations** have been defined as those with ARF incidence < 2:100000 school-age children or all age prevalence of RHD of < 1:1000 population per year
- ❑ Children not from low risk population have been considered to be at **moderate or high risk**

2) **Echocardiographic** evidence of **subclinical carditis** accepted as a major criteria (MR+/- AR)

# 2015 Revision of Jones Criteria

- 3) **Monoarthritis** has been included as a **major criteria** in **moderate or high risk** population
- 4) **Polyarthralgia** has been recognized as a major manifestation for **moderate or high risk population**
- 5) **Fever > 38.5 c**, **ESR > 60** for low risk population, and **fever > 38** and **ESR > 30** for moderate or high risk population

# 2015 Revision of Jones Criteria

## Low Risk Population

### Major criteria

- Carditis (clinical or subclinical)
- Arthritis (polyarthritis only)
- Chorea
- Erythema marginatum
- Subcutaneous nodule

### Minor criteria

- Polyarthralgia
- Fever ( $\geq 38.5$  °C)
- Elevation of ESR ( $\geq 60$  mm in the 1st hour) and/or CRP  $\geq 3$  mg/dL
- Prolonged PR interval, corrected for age (only when there is no carditis)

## Moderate to High Risk Population

### Major criteria

- Carditis (clinical or subclinical)
- Arthritis (polyarthritis, polyarthralgia, and/or monoarthritis)
- Chorea
- Erythema marginatum
- Subcutaneous nodule

### Minor criteria

- Fever ( $\geq 38.0$  °C)
- Elevation of ESR ( $\geq 30$  mm in the 1st hour) and/or CRP  $\geq 3$  mg/dL
- Prolonged PR interval, corrected for age (only when there is no carditis)

# 2015 Revised Jones Criteria

A firm diagnosis requires

1) 2 Major manifestations or 1 Major and 2 Minor manifestations

*and*

2) Evidence of a recent streptococcal infection.

# 2015 Revised Jones Criteria

Evidence of Preceding GAS Infection:

- 1) Increased or rising ASO titer or Anti-Dnase B titer
- 2) A positive throat culture

# RF Recurrences

- ❑ 2 major or 1 major and 2 minor or 3 minor manifestations for diagnosis
- ❑ Presence of antecedent streptococcal infection

# DDX of ARF

Presentation		
Polyarthrititis and fever	Carditis	Chorea
Septic arthritis (including disseminated gonococcal infection) <sup>†</sup>	Innocent murmur	Systemic lupus erythematosus
Connective tissue and other autoimmune disease <sup>††</sup>	Mitral valve prolapse	Drug intoxication
Viral arthropathy <sup>‡</sup>	Congenital heart disease	Wilson's disease
Reactive arthropathy <sup>‡</sup>	Infective endocarditis	Tic disorder <sup>‡</sup>
Lyme disease <sup>‡</sup>	Hypertrophic cardiomyopathy	Choreoathetoid cerebral palsy
Sickle cell anaemia	Myocarditis: viral or idiopathic	Encephalitis
Infective endocarditis	Pericarditis: viral or idiopathic	Familial chorea (including Huntington's)
Leukaemia or lymphoma		Intracranial tumour
Gout and pseudogout		Lyme disease <sup>‡</sup>
		Hormonal <sup>§</sup>



# Investigations

## Recommended for all cases

White blood cell count

Erythrocyte sedimentation rate (ESR)

C-reactive protein (CRP)

Blood cultures, if febrile

Electrocardiogram (if prolonged P-R interval or other rhythm abnormality, repeat in 2 weeks and again at 2 months, if still abnormal)

Chest X-ray, if clinical or echocardiographic evidence of carditis

**Echocardiogram** (consider repeating after 1 month, if negative)

Throat swab (preferably before giving antibiotics): culture for group A streptococcus

Antistreptococcal serology: both ASO and anti-DNase B titres, if available (repeat 10–14 days later if first test not confirmatory)

# Treatment of ARF

- ❑ Bed rest
- ❑ Salicylates: **Aspirin**
  - 60-100 mg/kg/day (maximum 8 g/day)  
given as 4 divided doses for 6-8 weeks
  - Attain a blood level 20-30 mg/dl
- ❑ Eradication of GAS from throat: BPG 1.2 MU im
- ❑ **Prednisolone**: 1-2 mg/kg/day taper over 6 weeks,  
taper gradually in severe carditis
- ❑ Heart Failure Treatment: **diuretics, ACEI**

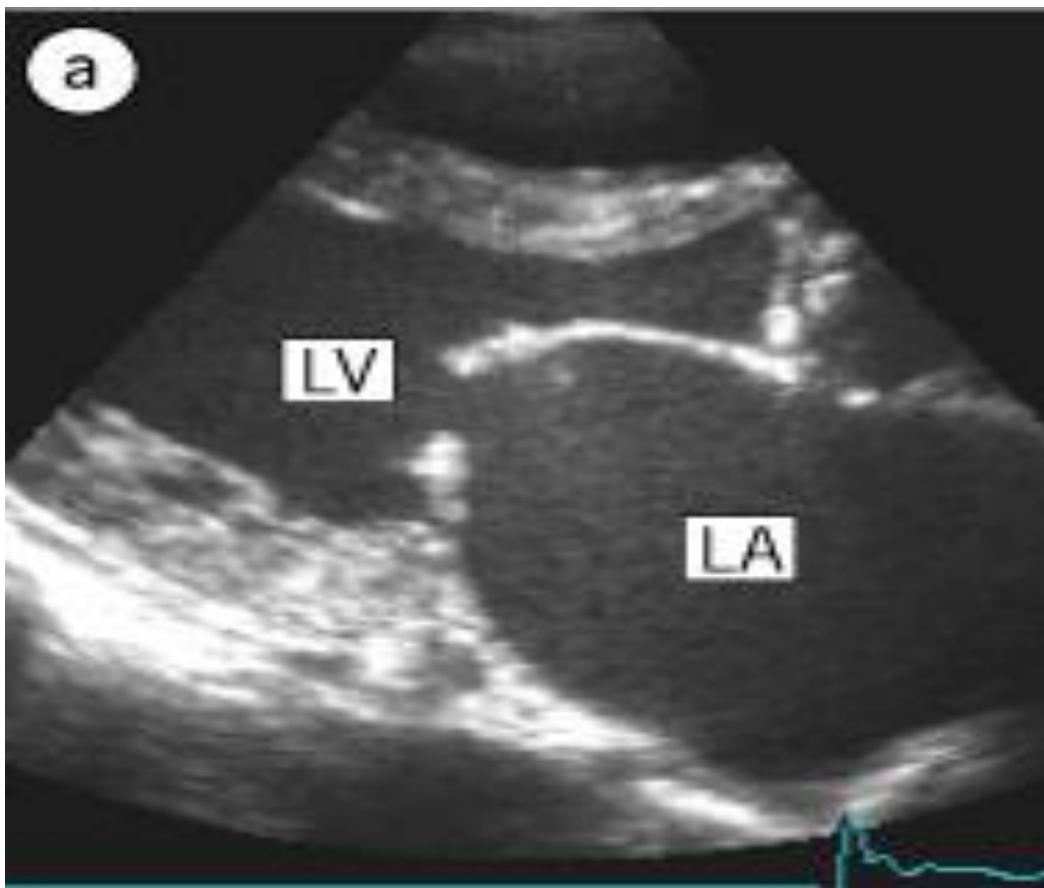
# Chronic Rheumatic Heart Disease

- ❑ Most commonly in Mitral-70%
- ❑ Frequently in Aortic-40%
- ❑ Less frequently Tricuspid-10%
- ❑ Rarely pulmonary valve-2%
- ❑ Mitral Stenosis is more common in females (3:1), while males have higher incidence of Aortic Regurgitation

# Mitral Stenosis

- ❑ The normal MVA = 4-5 cm<sup>2</sup>
  - In severe MS < 1.5 cm<sup>2</sup>
- ❑ High LAP
- ❑ The rise in LAP causes a similar rise in pulmonary capillaries, veins and pulmonary artery

# Mitral Stenosis



# Mitral Stenosis

## Symptoms

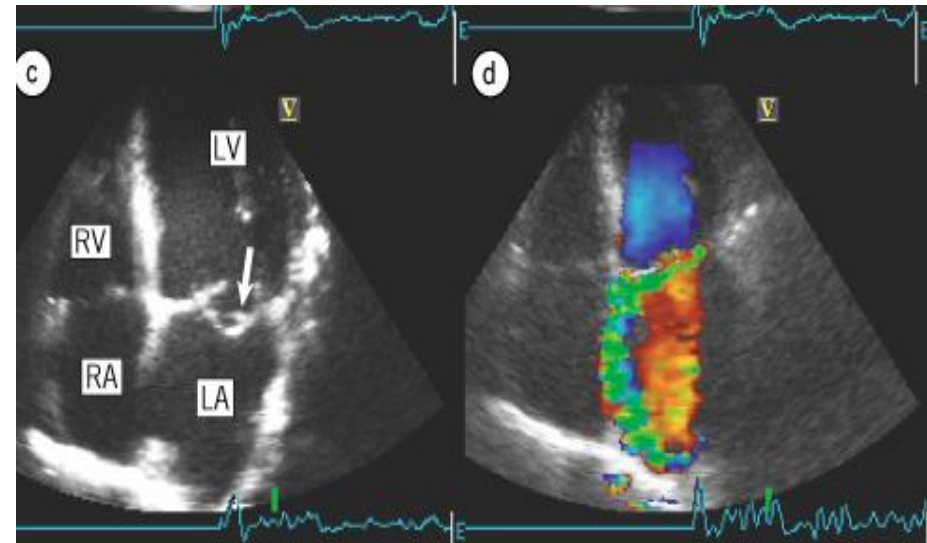
- ❑ Dyspnea
- ❑ Fatigue
- ❑ Palpitation
- ❑ Hemoptysis (10%)
- ❑ Hoarseness  
(Ortner's syndrome)
- ❑ Dysphagia
- ❑ Stroke or peripheral embolization

## Signs

- ❑ Cyanosis  
(Mitral facies, malar flush)
- ❑ Tapping apex ( S1)
- ❑ Parasternal heave
- ❑ Diastolic thrill
- ❑ Accentuated S1, accentuated S2
- ❑ Opening snap
- ❑ Mid-diastolic rumble

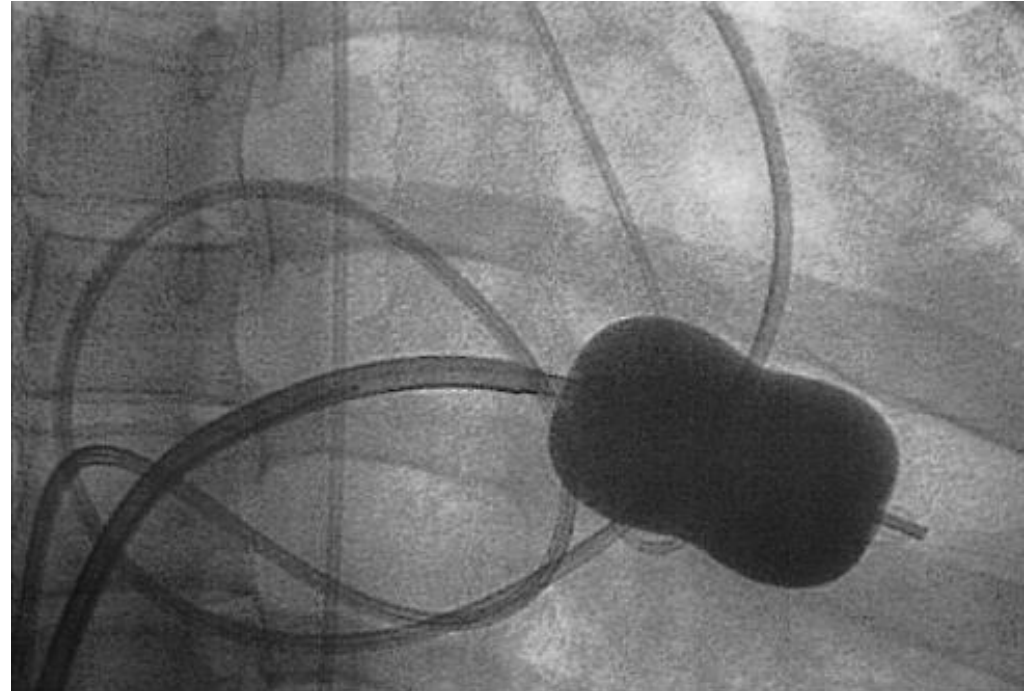
# Investigations

- ❑ CXR
  - Straightening of the left heart border
  - Double density
  - Kerley B lines , CA in MV
- ❑ ECG: LAE, P Mitrale, RV dominance
- ❑ Echodoppler



# Management

- ❑  $\beta$ -Blockers, CCB
- ❑ Digoxin (AF)
- ❑ Warfarin
- ❑ Balloon Valvuloplasty
- ❑ Mitral valve replacement





# Mitral Regurgitation

## Symptoms

- ❑ Asymptomatic
- ❑ Dyspnea, orthopnea, PND
- ❑ Displaced PMI, Thrill

## Signs

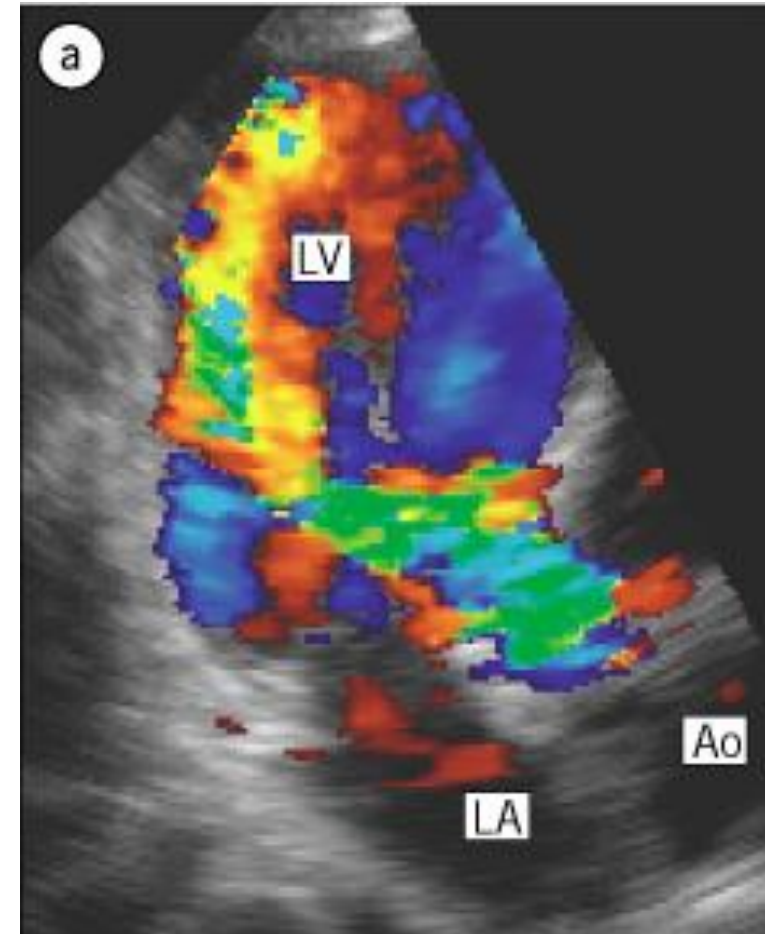
- ❑ Soft S1
- ❑ Pansystolic murmur
- ❑ Treatment is surgical

# Aortic Regurgitation

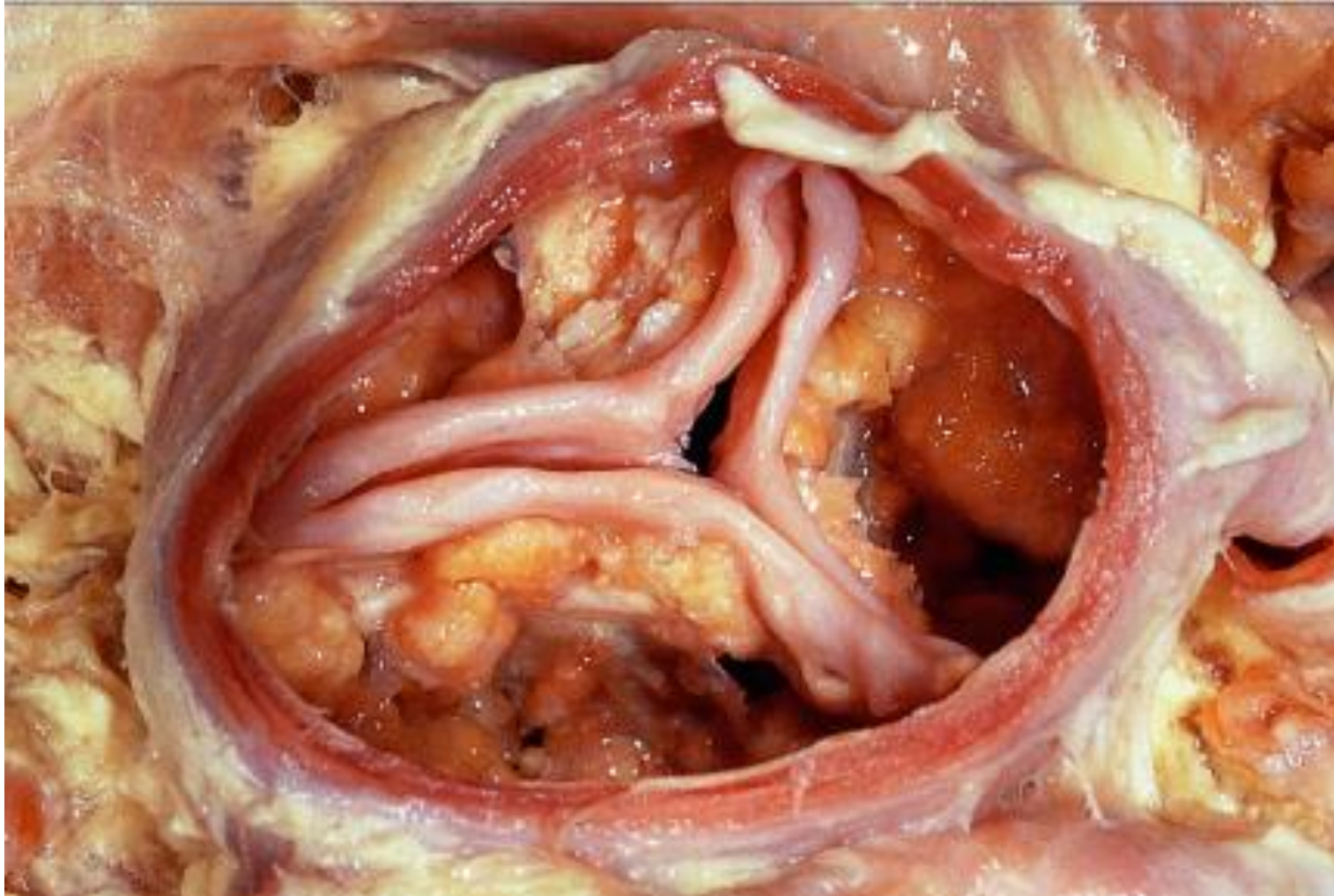
## Signs

- ❑ Water-hammer / collapsing pulse
- ❑ Wide pulse pressure
- ❑ Corrigan's sign
- ❑ De Musset sign
- ❑ Muller sign
- ❑ Quincke's pulse
- ❑ Hill's sign

## Echo



# Aortic Stenosis



# Aortic Stenosis

## Symptoms

- ❑ Angina
- ❑ Syncope
- ❑ Dyspnea

## Signs

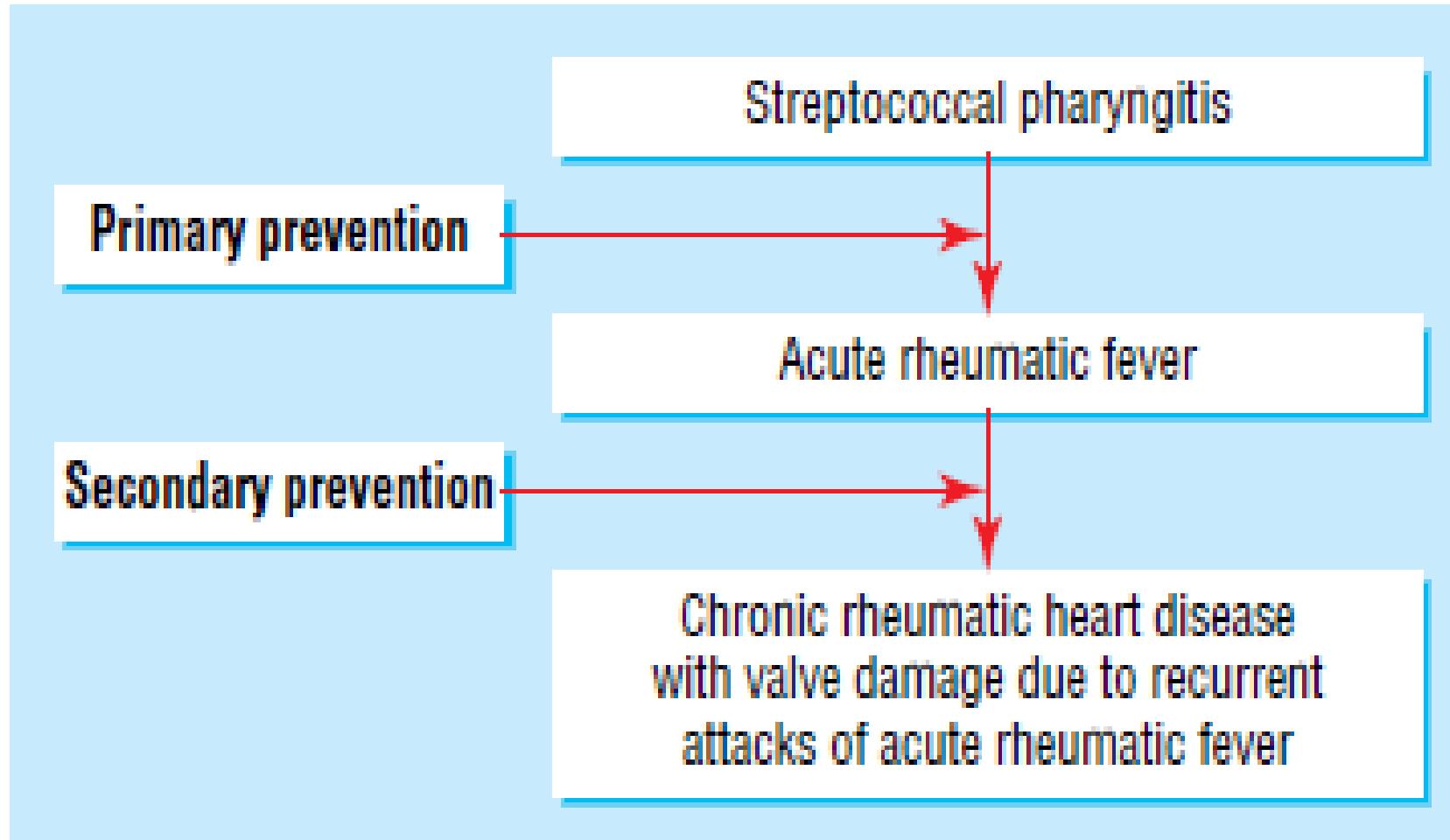
- ❑ Arterial Pulse wave form: Plateau
- ❑ Small (Parvus)
- ❑ Slow rise (Tardus)
- ❑ Sustained not displaced PMI
- ❑ Systolic thrill
- ❑ S4
- ❑ Late peaking of murmur
- ❑ Single S2: Soft or absent A2
- ❑ Paradoxical splitting of S2

# Aortic Stenosis

## Treatment

- ❑ Aortic valve Replacement
- ❑ Transcatheter Aortic Valve Replacement

# Prevention of ARF



# Secondary Prevention of RF

<b>Agent</b>	<b>Dose</b>	<b>Mode</b>
<b>Benzathine penicillin G</b>	<b>1 200 000 U every 4 weeks*</b>	<b>Intramuscular</b>
<b>or</b>		
<b>Penicillin V</b>	<b>250 mg twice daily</b>	<b>Oral</b>
<b>or</b>		
<b>Sulfadiazine</b>	<b>0.5 g once daily for patients 27 kg (60 lb) 1.0 g once daily for patients &gt;27 kg (60 lb)</b>	<b>Oral</b>
<b>For individuals allergic to penicillin and sulfadiazine</b>		
<b>Erythromycin</b>	<b>250 mg twice daily</b>	<b>Oral</b>

**\*In high-risk situations, administration every 3 weeks is justified and recommended**

# Duration of Secondary RF Prophylaxis

<b>Category</b>	<b>Duration</b>
<b>Rheumatic fever with carditis and residual heart disease (persistent valvular disease)</b>	<b>10 yrs since last episode or until age 40 yrs, (whichever is longer), sometimes life long prophylaxis</b>
<b>Rheumatic fever with carditis But no residual VHD</b>	<b>10 yrs or until age 21 yrs (whichever is longer)</b>
<b>Rheumatic fever without carditis</b>	<b>5 yrs or until age 21 yrs, (whichever is longer)</b>