Rheumatic Fever & Rheumatic Heart Disease

Dr. Abdulelah F Mobeirek FRCPC, FACC

Consultant Cardiologist at KFCC

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Lecture Outline

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

Definition

- **Acute rheumatic fever (ARF)** is a multisystem disease that follows **group A** β-hemolytic streptococcal throat infection
  - Represents a delayed immune response to infection with manifestations appearing after 2-4 weeks
  - Age 5-15 yrs
- **Rheumatic heart 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 M
- Annual incidence of RF = 0.5 M → nearly ½ develop carditis
- Estimated deaths from RHD = 250,000 / year
- Imposes a substantial burden on health care systems with limited budgets

Epidemiologic Background

- The incidence of RF & the prevalence of RHD has declined substantially in Europe, North America and other developed nations
  - This decline has ben attributed to improved hygiene, reduced household crowding, and improved medical care
- The major burden is currently found in low & middle-income countries (India, Middle East, Gulf countries), and in selected indigenous populations of certain developed countries (Australia & New Zealand).
- A disease of poverty & low socioeconomic status
- In underdeveloped countries RHD is the leading cause of CV death during the first 5 decades of life

ARF & RHD in Saudi Arabia

- Published data in KSA is limited.
- In developed countries the incidence of ARF has declined over past 50 y, incidence ranging 0.2–0.64 / 100,000 (USA).
- ARF incidence in Eastern province was 22 / 100,000, age 5-14 y.
- 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 & signs are shared by many inflammatory & infectious diseases
- Accurate diagnosis is important
- Overdiagnosis will result in individuals receiving treatment unnecessarily
- Underdiagnosis may lead to further episodes of ARF causing damage, the need for valve surgery, CHF and death

Diagnosis of ARF

- Diagnosis is primarily clinical and is based on a constellation of signs & 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 (detected by echo)
- Murmurs of MR or AR may occur in acute stage while mitral stenosis occurs in late stages
- Signs of carditis: tachycardia, murmurs of MR/AR, cardiomegay and signs of CHF

Arthritis

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

Sydenham Chorea

- Also known as Saint Vitus' dance
- Occur in 10-15%, extrapyramidal manifestation, female predominance
- Abrupt purposeless involuantry movements of muscles of face, neck, trunk, & limbs.
- Can be a delayed manifestation of ARF - months
- Clinically manifests 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 & in 2015

2015 Revision of Jones Criteria

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 & ESR > 30 for moderate or high-risk population**

2015 Revised Jones Criteria

Low-risk Population

Major criteria

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

Minor criteria

- Polyarthralgia
- Fever (≥38.5 °C)
- Elevation of ESR (≥60 mm in the 1st hour) and/or CRP ≥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 (≥38.0 °C)
- Elevation of ESR (≥30 mm in the 1st hour) and/or CRP ≥3 mg/dL
- Prolonged PR interval, corrected for age (only when there is no carditis)

2015 Revised Jones Criteria

A firm diagnosis of initial ARF attack requires

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

   and

2) Evidence of a recent streptococcal infection.
   - Increased or rising ASO titer or Anti-DNAse B titer
   - A positive throat culture

ARF Recurrences

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

Differential Diagnosis of ARF

Presentation		
Polyarthritis and fever	**Carditis**	**Chorea**
Septic arthritis (including disseminated gonococcal infection)†	Innocent murmur	Systemic lupus erythematosus
Connective tissue and other autoimmune disease††	Mitral valve prolapse	Drug intoxication
Viral arthropathy¥	Congenital heart disease	Wilson's disease
Reactive arthropathy¥	Infective endocarditis	Tic disorder‡
Lyme disease≠	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≠
		Hormonal§

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^2
  - In severe MS < 1.5 cm^2
- High LAP
- The rise in LAP causes a similar rise in pulmonary capillaries, veins & pulmonary artery

Mitral Stenosis

Symptoms

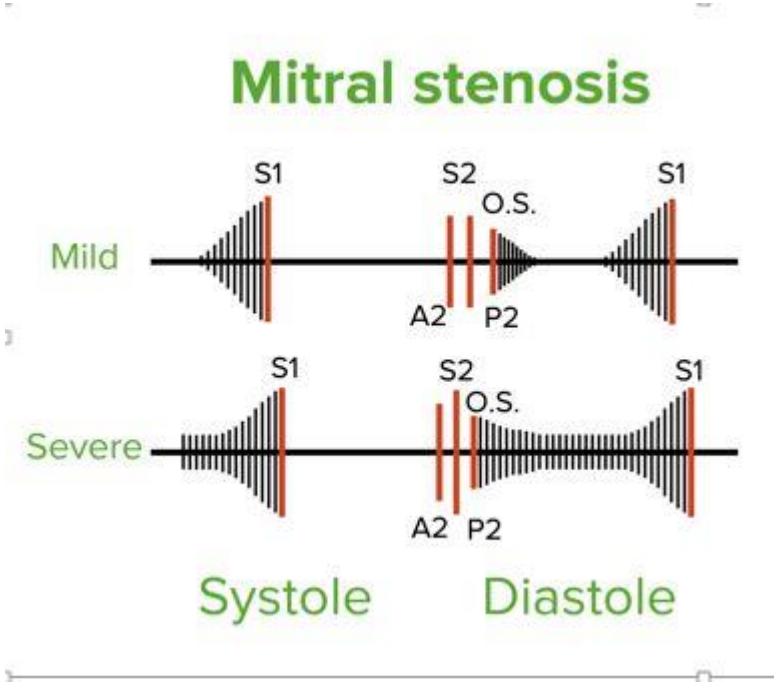
- Dyspnea
- Fatigue
- Palpitation
- Hemoptysis
- Hoarseness (Ortner's syndrome)
- Dysphagia
- Stroke or peripheral embolization

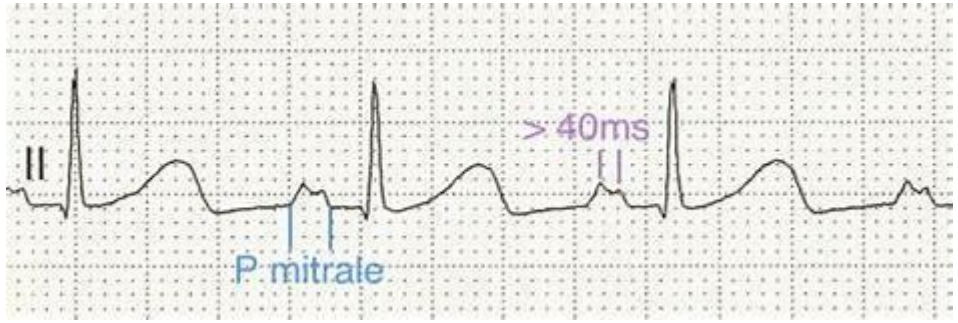
Signs

- 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


Mitral stenosis
S1
S2
O.S.
S1
Mild
A2
P2
S1
S2
O.S.
S1
Severe
A2
P2
Systole
Diastole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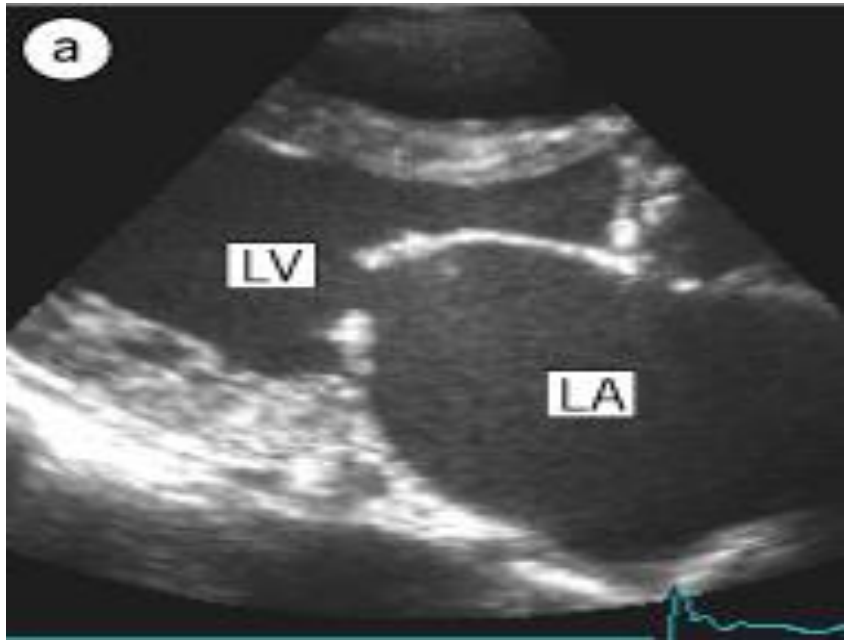
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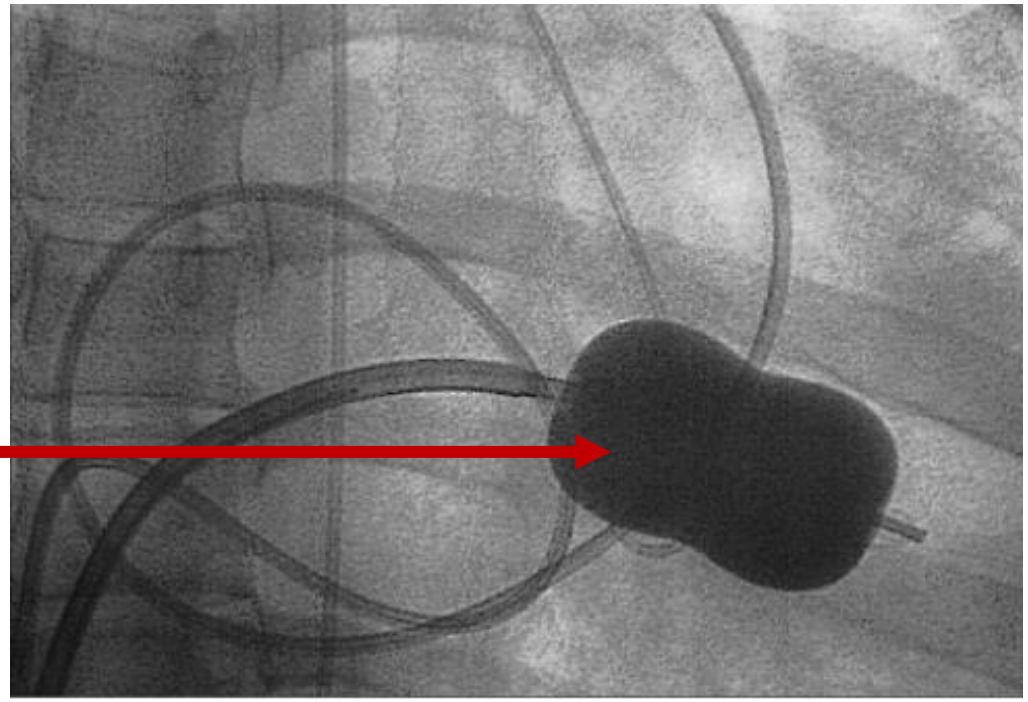
LA

Management

- β-Blockers, CCB
- Digoxin (AF)
- Warfarin
- Balloon valvuloplasty
- Mitral valve replacement (MVR)



Mitral Regurgitation

Symptoms

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

Signs

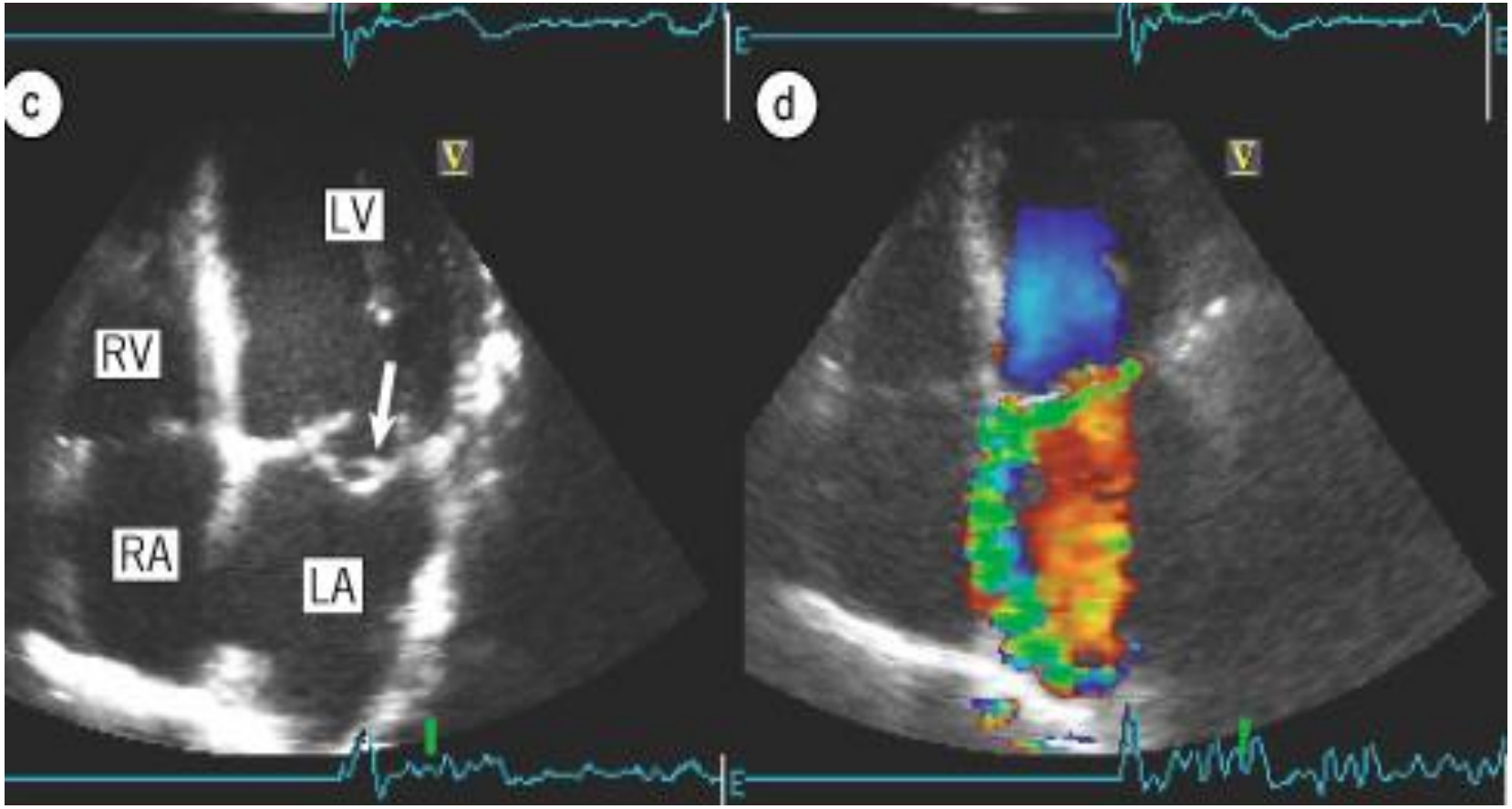
- Soft S1
- Pansystolic murmur

Treatment

- Surgical - MVR

Mitral Regurgitation

Echo



Aortic Regurgitation

Symptoms

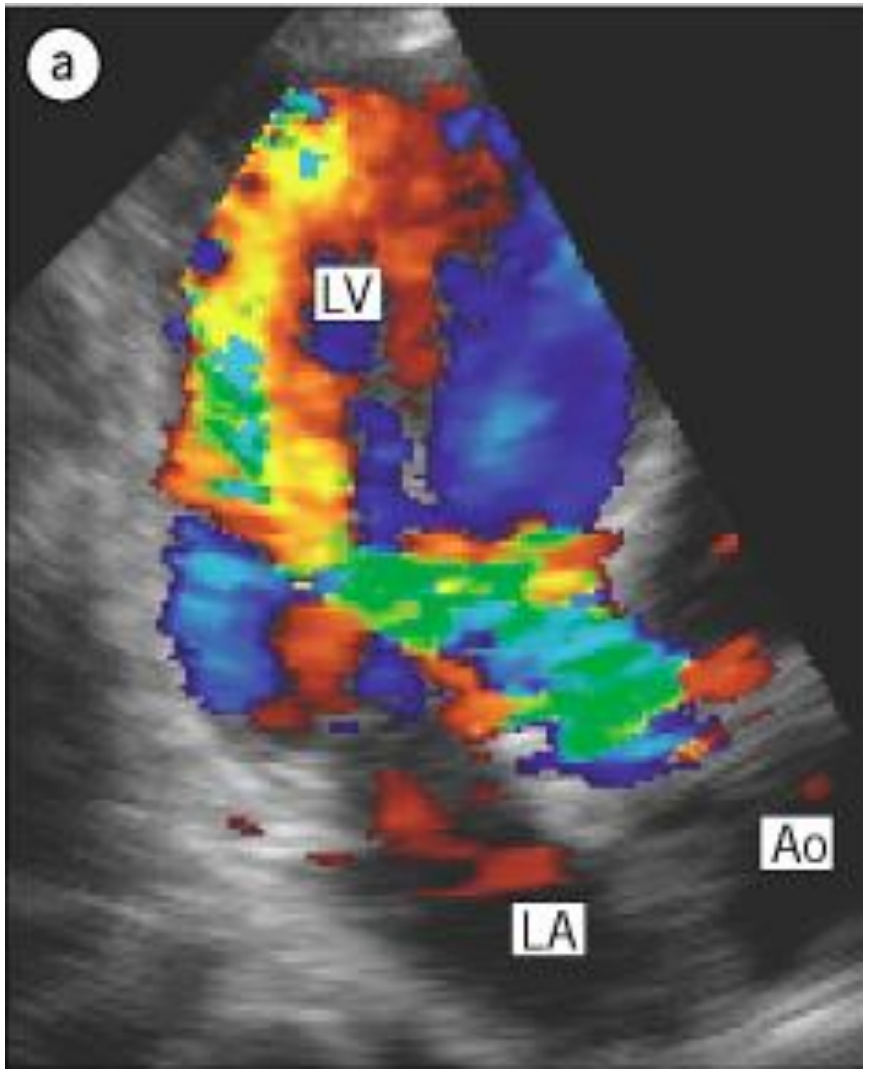
- Shortness of breath on exertion (SOBE)

Signs

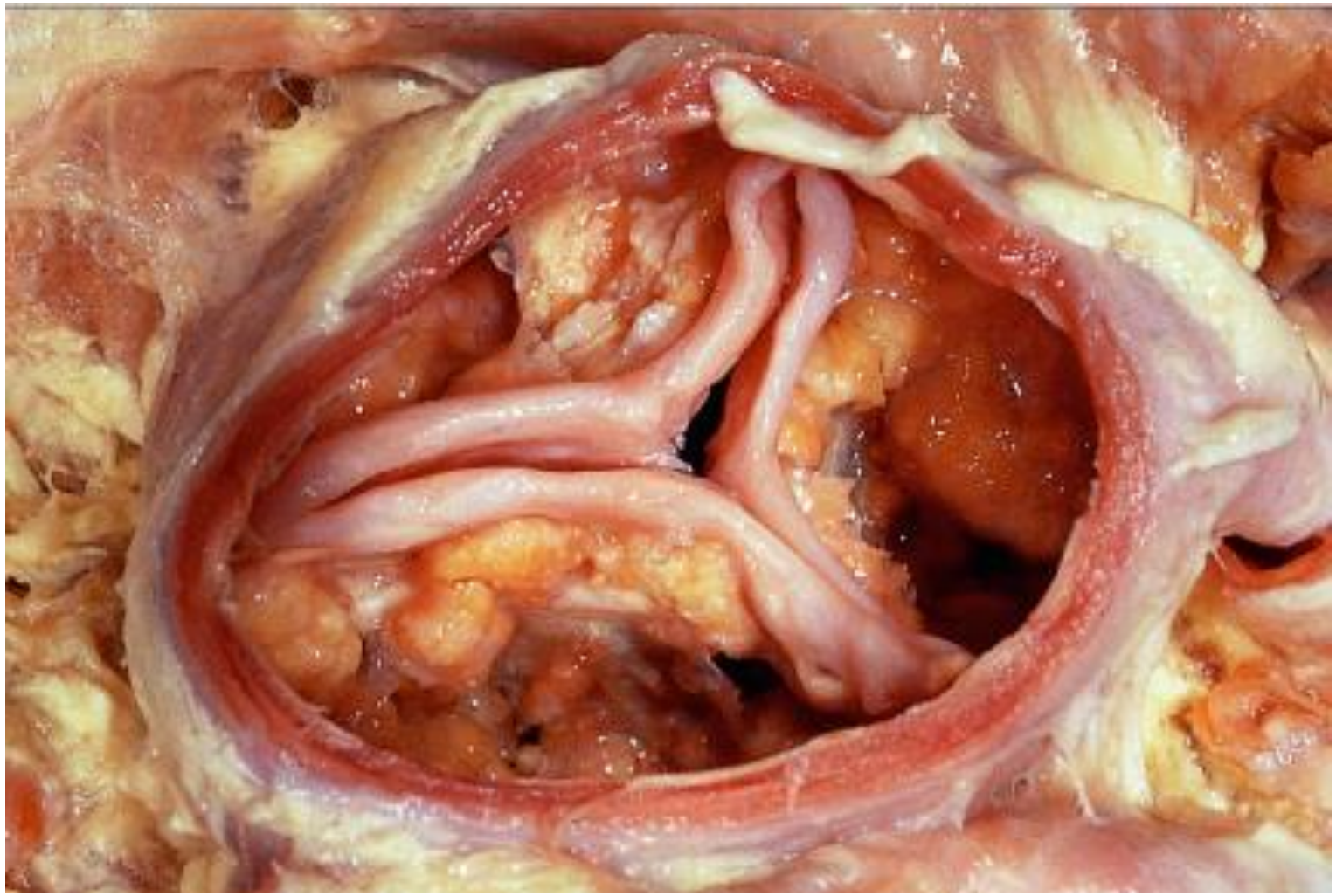
- Water-hammer / collapsing pulse
- Wide pulse pressure
- Corrigan's sign
- De Musset sign
- Muller's sign
- Quincke's pulse
- Hill's sign
- Traube's sign
- Duroziez sign
- Early diastolic murmur
- Austin-Flint murmur

Aortic Regurgitation

Echo



Aortic Stenosis



Aortic Stenosis

Symptoms

- Angina
- Syncope
- Dyspnea

Signs

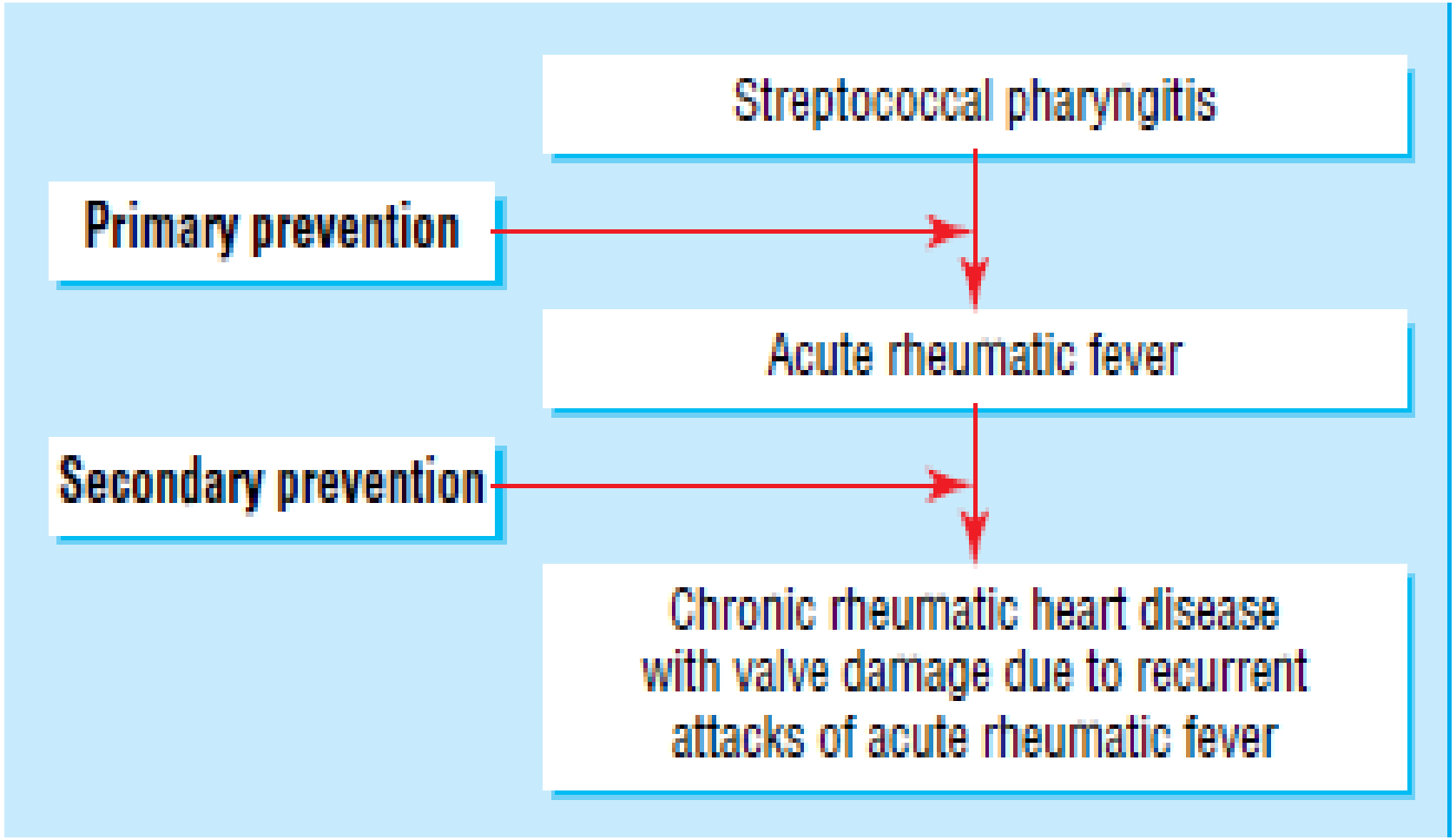
- Arterial pulse wave form: Plateau
- Small (Parvus)
- Slow rise (Tardus)
- Sustained not displaced PMI
- Systolic thrill
- S4
- Late peaking of murmur
- Single S2: soft or absent A2
- Paradoxical splitting of S2

Aortic Stenosis

Treatment

- Aortic valve Replacement
- Transcatheter Aortic Valve Replacement

Prevention of ARF



Secondary Prevention of RF

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

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

Duration of Secondary RF Prophylaxis

Category	Duration
Rheumatic fever with carditis & residual heart disease (persistent valvular disease)	**10 y since last episode or until age 40 y, (which-ever is longer), sometimes life-long prophylaxis**
Rheumatic fever with carditis	**10 yrs or until age 21 y**
But no residual VHD	**(whichever is longer)**
Rheumatic fever without carditis	**5 y or until age 21 y, (whichever is longer)**