

# Acute Rheumatic Fever

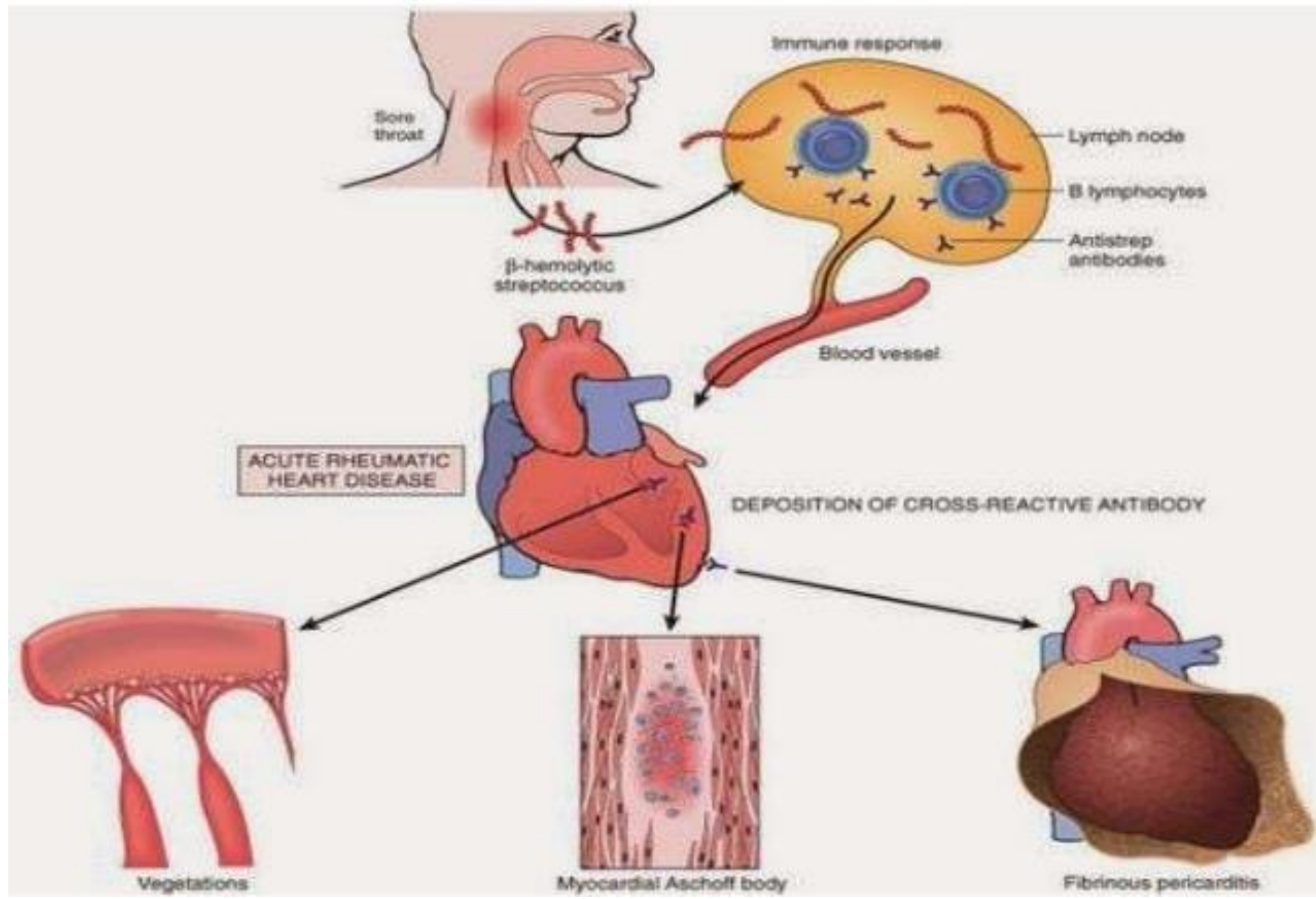
By:

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Acute rheumatic fever usually affects children and young adults between the ages of 5 and 15 years.

## **Pathogenesis**

The condition is triggered by an immune-mediated delayed response to infection with specific strains of **group A streptococci**, which have antigens that cross-react with cardiac myosin and sarcolemmal membrane proteins.



## **Clinical features**

It is a multisystem disorder

It usually presents with fever, anorexia, lethargy and joint pain, 2–3 weeks after an episode of streptococcal pharyngitis.

## Carditis

Rheumatic fever causes a pancarditis involving the endocardium, myocardium and pericardium to varying degrees.

## ARTHRITIS

This is the most common major manifestation

- Acute
- Painful
- Asymmetric
- migratory inflammation
- of the large joints typically affects the knees, ankles, elbows and wrists.
- Dramatic response to NSAID

## Skin lesions

**Erythema marginatum** occurs in less than 5% of patients.

- start as red macules that fade in the centre but remained at the edges
- occur mainly on the trunk and proximal extremities but not the face.

**Subcutaneous nodules** occur in 5–7% of patients.

- small (0.5–2.0 cm), firm and painless
- best felt over extensor surfaces of bone or tendons
- appear more than 3 weeks after the onset of other manifestations and therefore help to confirm rather than make the diagnosis.

# Rheumatic fever-diagnosis



Subcutaneous nodules  
*(nodules of rheumatoid arthritis are larger)*



# Erythema marginatum / Subcutaneous nodules

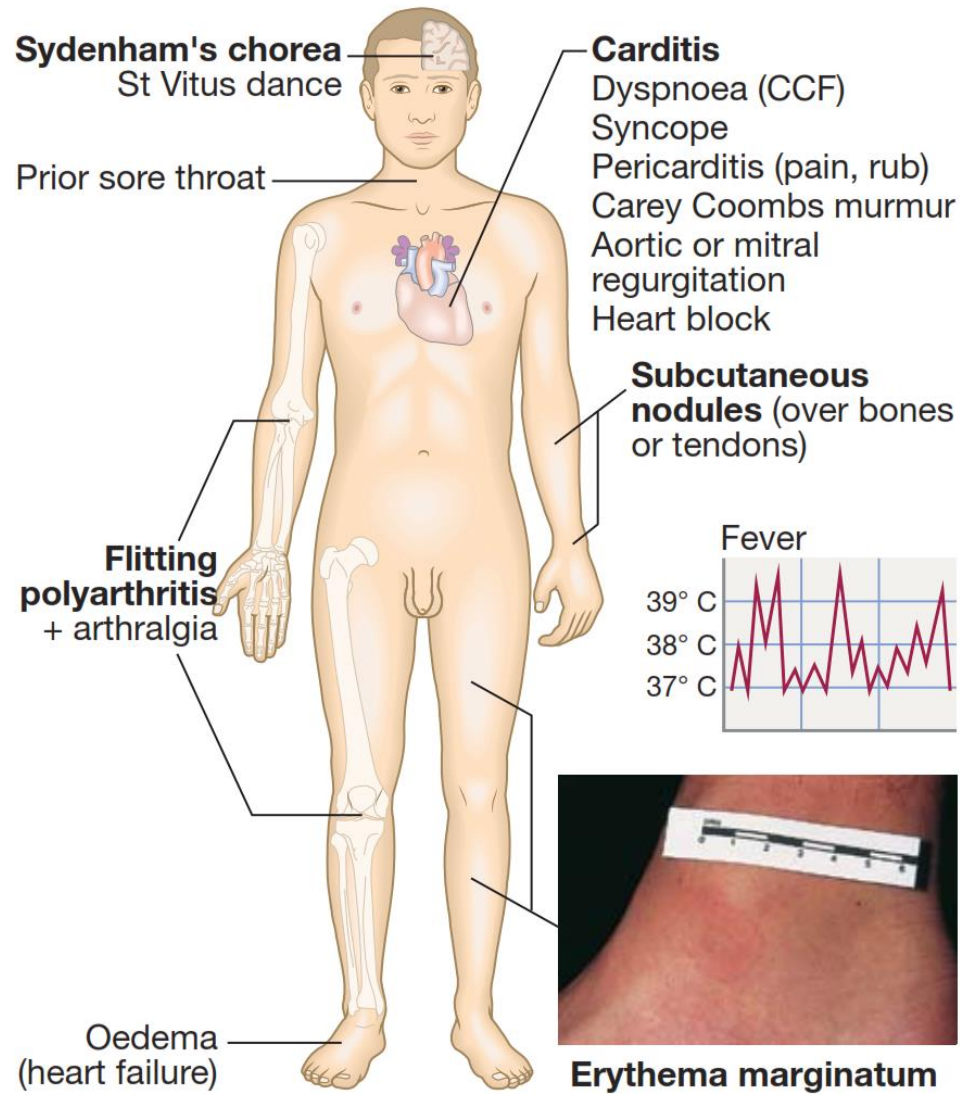


<http://www.hxbenefit.com/erythema-marginatum.htm>

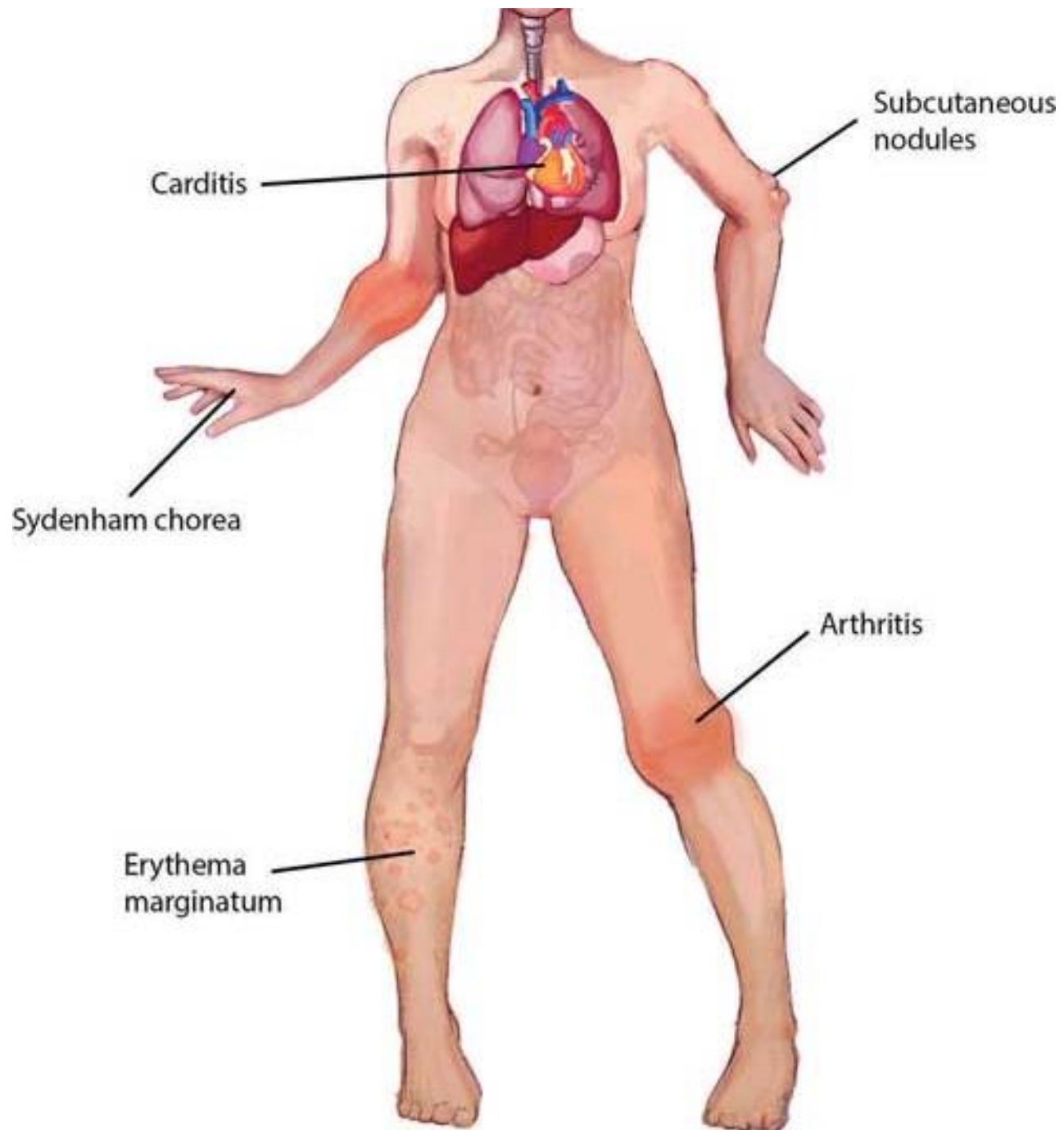
<http://www.doctortipster.com/1789-rheumatic-fever.html>

## Sydenham's chorea

- also known as St Vitus dance
- It is a late neurological manifestation that appears at least 3 months after the episode of acute rheumatic fever, when all the other signs may have disappeared.
- It occurs in up to one-third of cases
- It is more common in females.
- It is a purposeless, involuntary, choreiform movements of the hands, feet or face.
- Emotional lability may be the first feature
- Spontaneous recovery usually occurs within a few months.



**Fig. 16.80** Clinical features of rheumatic fever. Bold labels indicate Jones major criteria. (CCF = congestive cardiac failure) *Inset (Erythema marginatum)* From Savin JA, Hunter JAA, Hepburn NC. *Skin signs in clinical medicine*. London: Mosby-Wolfe, Elsevier; 1997.



## Other rare features

- Pleurisy
- pleural effusion
- pneumonia

# Investigations

- WBC, ESR and CRP : for monitoring progress of the disease
- Throat cultures :positive results are obtained in only 10–25%
- Antistreptolysin O antibodies (ASO) : normal in one-fifth of adult cases of rheumatic fever and normal most cases of chorea.
- Echocardiography should be carried out and typically shows mitral regurgitation ; aortic regurgitation and pericardial effusion.

**i****16.76 Investigations in acute rheumatic fever****Evidence of a systemic illness**

- Leucocytosis, raised erythrocyte sedimentation rate and C-reactive protein

**Evidence of preceding streptococcal infection**

- Throat swab culture: group A  $\beta$ -haemolytic streptococci (also from family members and contacts)
- Antistreptolysin O antibodies (ASO titres): rising titres, or levels of >200 U (adults) or >300 U (children)

**Evidence of carditis**

- Chest X-ray: cardiomegaly; pulmonary congestion
- ECG: first- and, rarely, second-degree atrioventricular block; features of pericarditis; T-wave inversion; reduction in QRS voltages
- Echocardiography: cardiac dilatation and valve abnormalities

# i 16.75 Jones criteria for the diagnosis of rheumatic fever

## Major manifestations

- Carditis
- Polyarthritis
- Chorea
- Erythema marginatum
- Subcutaneous nodules

## Minor manifestations

- Fever
- Arthralgia
- Raised erythrocyte sedimentation rate or C-reactive protein
- Previous rheumatic fever
- Leucocytosis
- First-degree atrioventricular block

## Plus

- Supporting evidence of preceding streptococcal infection: recent scarlet fever, raised antistreptolysin O or other streptococcal antibody titre, positive throat culture\*

\*Evidence of recent streptococcal infection is particularly important if there is only one major manifestation.

## For all patient populations with evidence of preceding GAS infection

Diagnosis: Initial ARF	2 major manifestations or 1 major plus 2 minor manifestations
Diagnosis: Recurrent ARF	2 major or 1 major and 2 minor or 3 minor

A presumptive diagnosis can be made **without evidence of preceding streptococcal infection** in cases of isolated **chorea or carditis**, if other causes of these have been excluded



# Management

The aims

- limit cardiac damage
- relieve symptoms.

## Bed rest

- The duration should be guided by symptoms, along with temperature, leucocyte count and ESR
- Patients can then return to normal physical activity but strenuous exercise should be avoided in those who have had carditis.

## Treatment of cardiac failure

Some patients, particularly those in early adolescence, can develop a fulminant form of the disease with severe mitral regurgitation and, sometimes, concomitant aortic regurgitation.

If heart failure in these cases **does not respond to medical treatment, valve replacement may be necessary**

Occasionally, AV block may occur but is seldom progressive and usually resolves spontaneously.

Rarely, pacemaker insertion may be required.

## Acute Antibiotics treatment

A single dose of benzathine benzylpenicillin (1.2 million U IM)

or

oral phenoxymethylpenicillin (250 mg 4 times daily for 10 days)

If the patient is penicillin-allergic, erythromycin or a cephalosporin can be used.

## long-term prophylaxis

oral phenoxymethylpenicillin (250 mg twice daily)

or

benzathine benzylpenicillin (1.2 million U IM monthly), if adherence is in doubt.

Sulfadiazine or erythromycin may be used if the patient is allergic to penicillin

## Antibiotic prophylaxis can usually be stopped :

### Without residual heart disease :

prophylaxis should continue until 5 years after the last episode or 21 years of age, whichever is later.

### With residual heart disease:

prophylaxis should continue until 10 years after the last episode or 40 years of age, whichever is later.

But the duration of prophylaxis should be extended if the patient lives in an area of high prevalence and has an occupation (such as teaching) with a high risk of exposure to streptococcal infection.

NOTE: While long-term antibiotic prophylaxis prevents further attacks of acute rheumatic fever, it does not protect against infective endocarditis

## Aspirin

- a response within 24 hours helps confirm the diagnosis.
- A reasonable starting dose is 60 mg/kg body weight/day, divided into six doses. In adults, 100 mg/kg per day may be needed up to the limits of tolerance or a maximum of **8 g per day**.
- Aspirin should be continued until the ESR has fallen and then gradually tailed off.

## Glucocorticoids

- These produce more rapid symptomatic relief than aspirin and
- are indicated in cases with **carditis or severe arthritis**.
- There is no evidence that long-term steroids are beneficial.
- Prednisolone (1.0–2.0 mg/kg per day in divided doses) should be continued until the ESR is normal and then tailed off.

## Chronic rheumatic heart disease

Chronic valvular heart disease develops in at **least half of those** affected by rheumatic fever with carditis.

Two-thirds of cases occur in **women**.

a history of rheumatic fever or chorea in only **about half of all patients** with chronic rheumatic heart disease.

**The mitral valve is affected in more than 90% of cases**

The aortic valve is the next most frequently involved followed by the tricuspid and then the pulmonary valve.

Isolated mitral stenosis accounts for about 25% of all cases

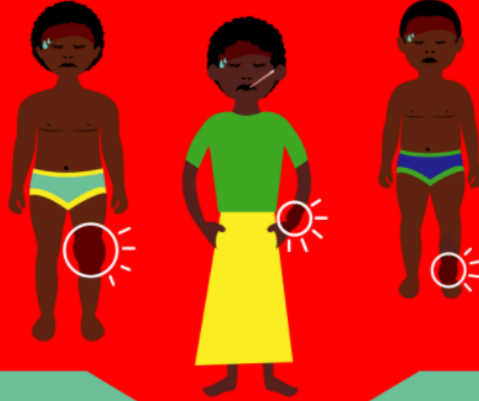
**an additional 40% have mixed mitral stenosis and regurgitation.**

# JOINT PAIN + FEVER CAN BE: RHEUMATIC FEVER

Malaria is not the only cause of joint pain and fever.

Joint pain and fever can indicate

**Acute Rheumatic Fever.**



Two atipa pee n keken aye kelo arem me wang mwot kede lyeto ento twero bedo anyut me **two adunu nyo two itao.**

**Acute Rheumatic Fever can damage the heart.**



**Two adunu nyo two itao twero balo adunu ni woko.**

If your child **(3-17 yrs)** has joint pain and fever take them to the nearest health center for evaluation.



Cwal atini **(me mwaka 3 nio 18)** matye kede arem me wang mwot kede lyeto l ot yat ma cok kedi wek opim.

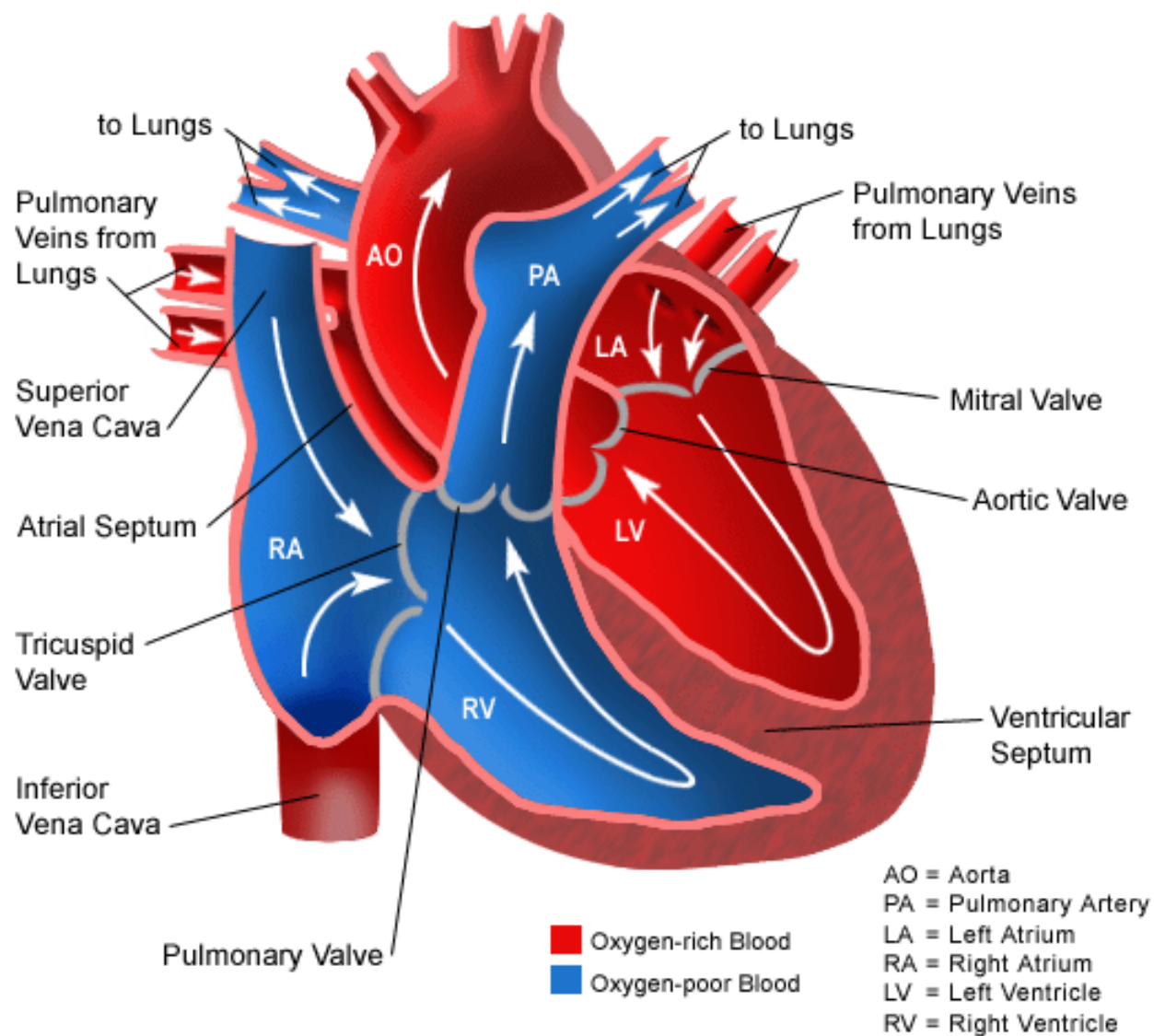
**FREE EVALUATIONS IN LIRA REGIONAL REFERRAL HOSPITAL**

Dakatal adwong me Lira  
TYE ME NONO



# **Valvular heart disease**

## Normal Heart



LVEDP	Left ventricle end diastolic pressure
PVH	pulmonary venous hypertension
PAH	pulmonary arterial hypertension
RVH	Right ventricular hypertrophy
RV	Right ventricle
TR	Tricuspid regurgitation
RA	Right atrium
CHF	congestive heart failure

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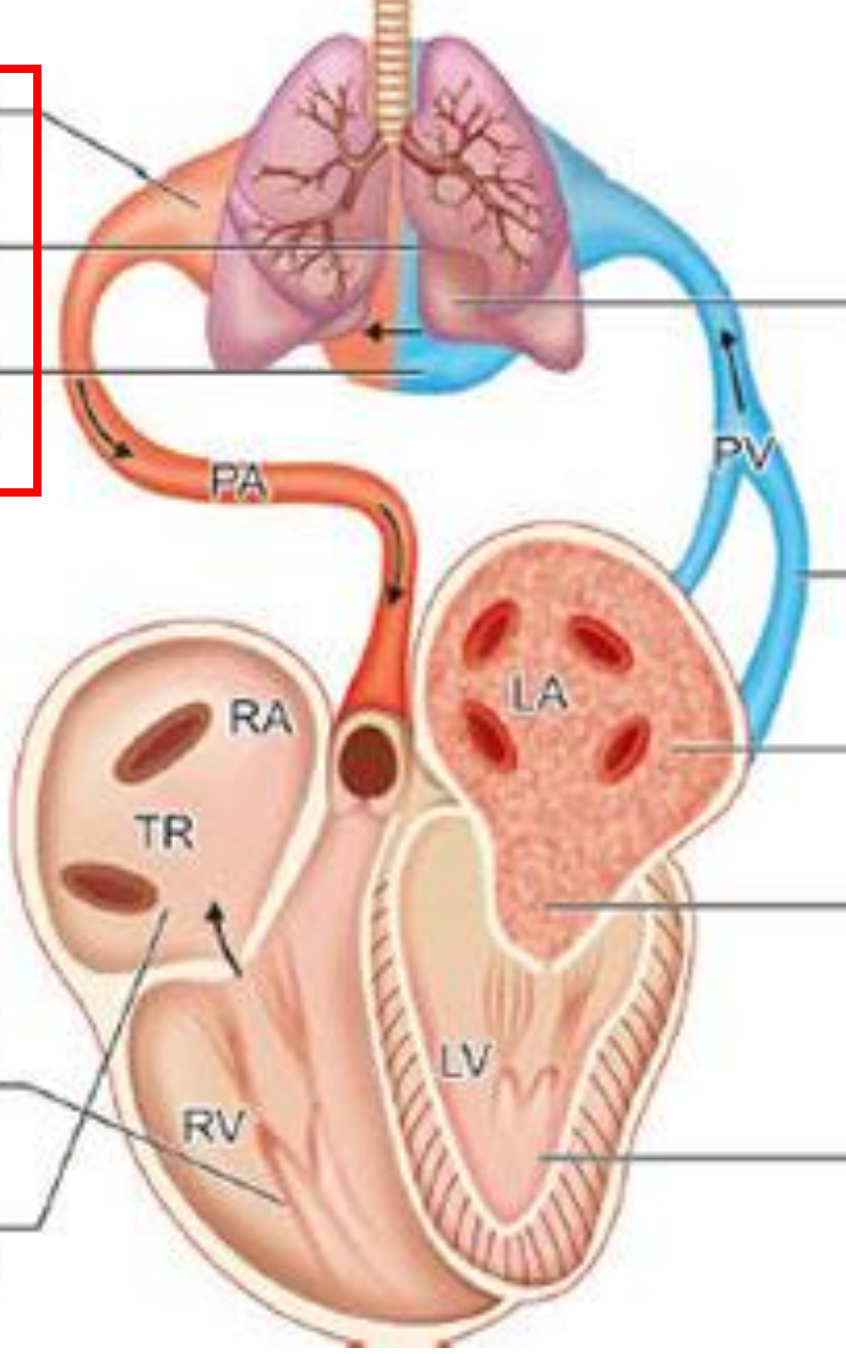
Pulmonary artery vasoconstriction  
 Interstitial edema  
 Pulmonary vascular changes

5

PAH

6

RVH followed by RV dilatation  
 TR and raised RA pressure—CHF



Passive transmission of LAP

3

PVH

2

Elevated LA pressure

1

Mitral valve disease

1

Elevated LVEDP

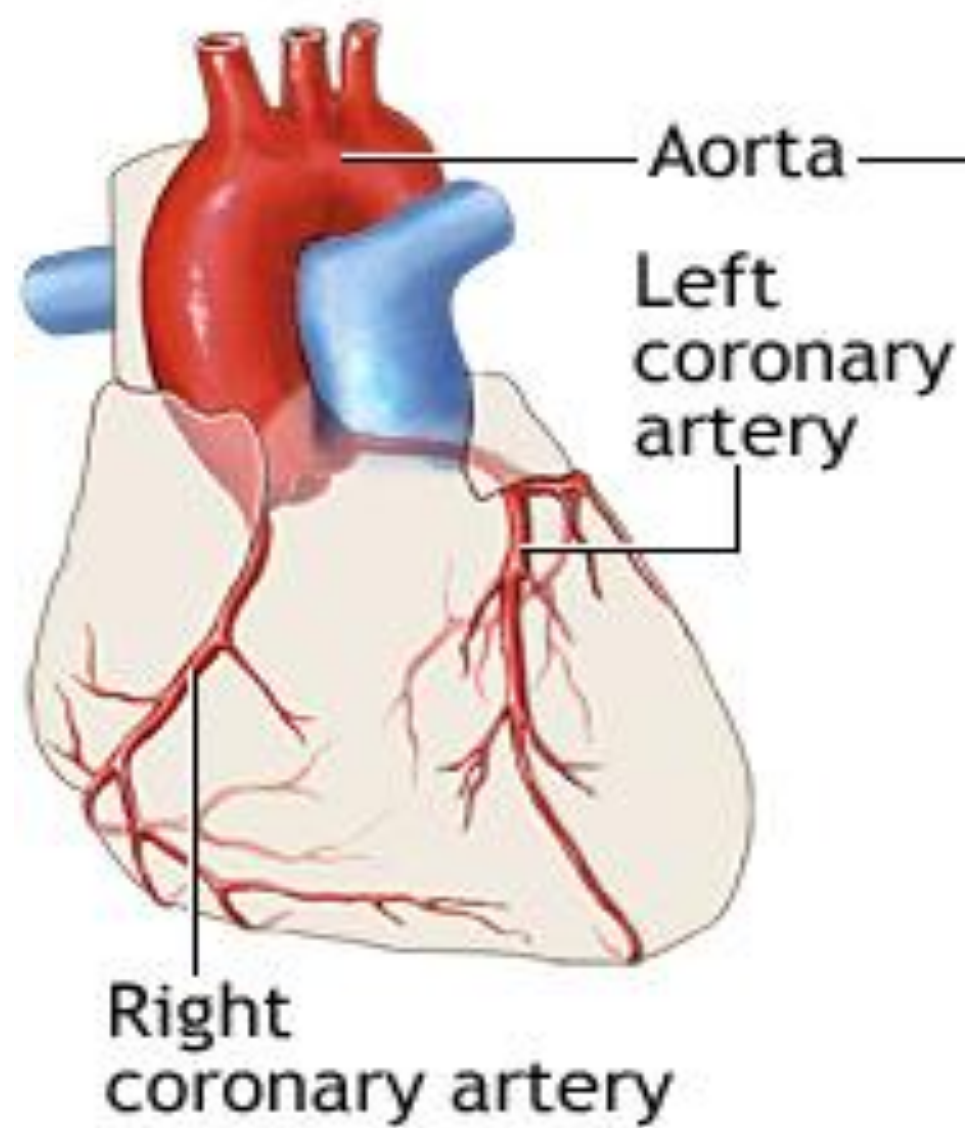
Aortic valve disease

## Valvular heart disease

### Symptoms:

- **Asymptomatic**
- **Chest pain :** Aortic stenosis , Pulmonary hypertension
- **Palpitation :** Arrhythmias ( atrial fibrillation )
- **Syncope :** Low cardiac output ( severe stenosis )
- **Fatigue :** Low cardiac output
- **Oedema :** Right sided heart failure
- **Breathlessness :** Pulmonary congestion
- **Cough :** Pulmonary congestion
- **Haemoptysis :** Pulmonary congestion
- **Thromboembolism :** Atrial fibrillation
- **Sudden death :** severe aortic stenosis , arrhythmias

## Normal heart



## Signs:

- **Mitral facies : mitral stenosis**
- **Abnormal pulse : large volume collapsing ( aortic regurgitation ) , slow rising ( aortic stenosis )**
- **Displaced apex beat : cardiomegaly**
- **Abnormal heart sounds:**
  - Loud S1 : mitral stenosis**
  - Soft S2 : Mitral regurgitation**
  - Soft S2 : Aortic stenosis**
  - Loud P2 : Pulmonary hypertension**
  - S3 : Mitral regurgitation , left heart failure**
  - S4 : Aortic stenosis**
- **Murmur**
- **Thrill : severe mitral regurgitation , severe aortic stenosis**
- **Heave : Pulmonary hypertension**
- **Crepitation : pulmonary odema ( left sided heart failure )**
- **Oedema ( ankle oedema , ascites , pleural effusion , hepatic congestion ): Right sided heart failure**

# Mitral Facies

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# ***Investigations :***

- ***ECG :*** ***chambers enlargement , arrhythmias***
- ***Chest X ray :*** ***chambers enlargement , pulmonary congestion***
- ***Echocardiography :*** ***valves , cardiac chambers size and /or function***
- ***Cardiac catheterization :*** ***coexistent coronary artery diseases , severity of valvular disease***



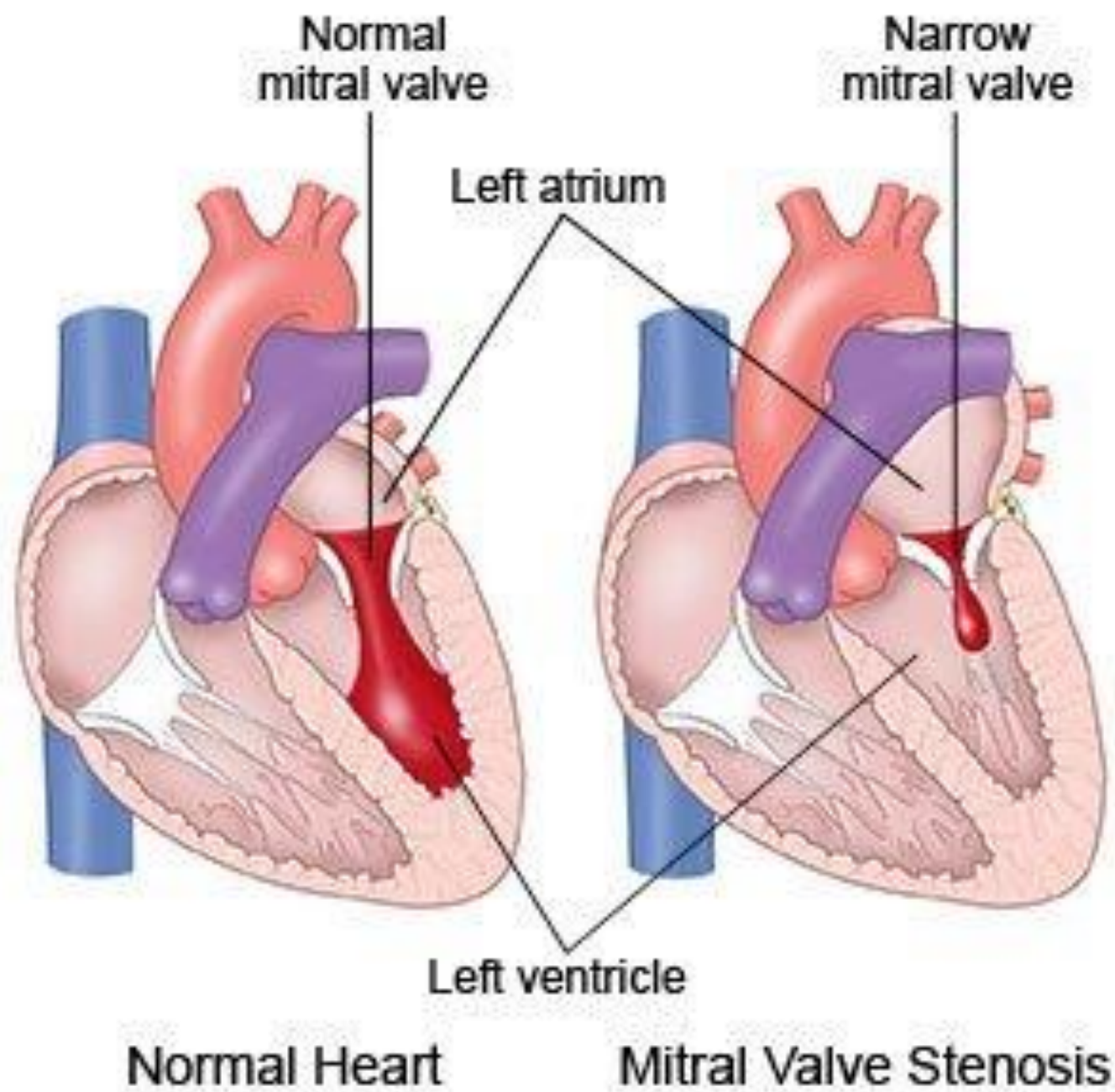
## Management:

Patients with asymptomatic and mild valvular disease can be treated medically

- Symptomatic patients and/or severe valvular disease : intervention by balloon valvuloplasty ( stenosis ) , valve repair ( regurgitation ) or valve replacement should be considered
- When aortic root dilatation is the cause of aortic regurgitation, as can occur in Marfan's syndrome, aortic root replacement is usually necessary.

## Medical management

- anticoagulation in atrial fibrillation
- ventricular rate control with digoxin, B blockers or rate-limiting calcium channel blockers in atrial fibrillation
- Diuretic to control pulmonary congestion.
- Vasodilators like ACEI or ARB if systemic hypertension ( cautious in severe aortic stenosis )
- Treatment may be required for underlying conditions, such as endocarditis



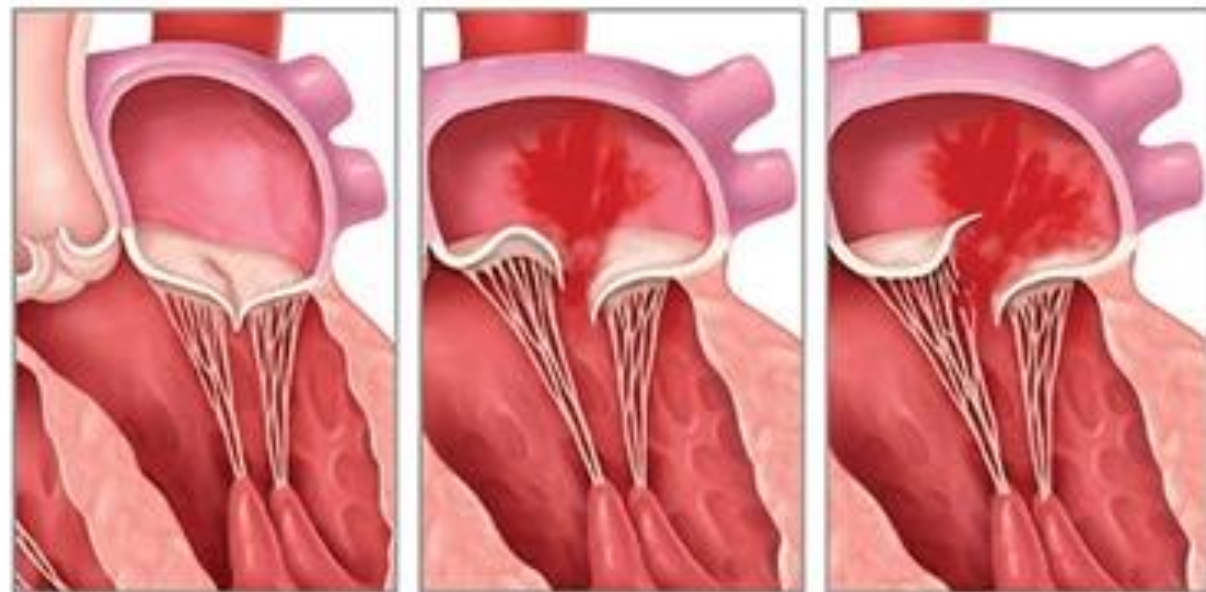
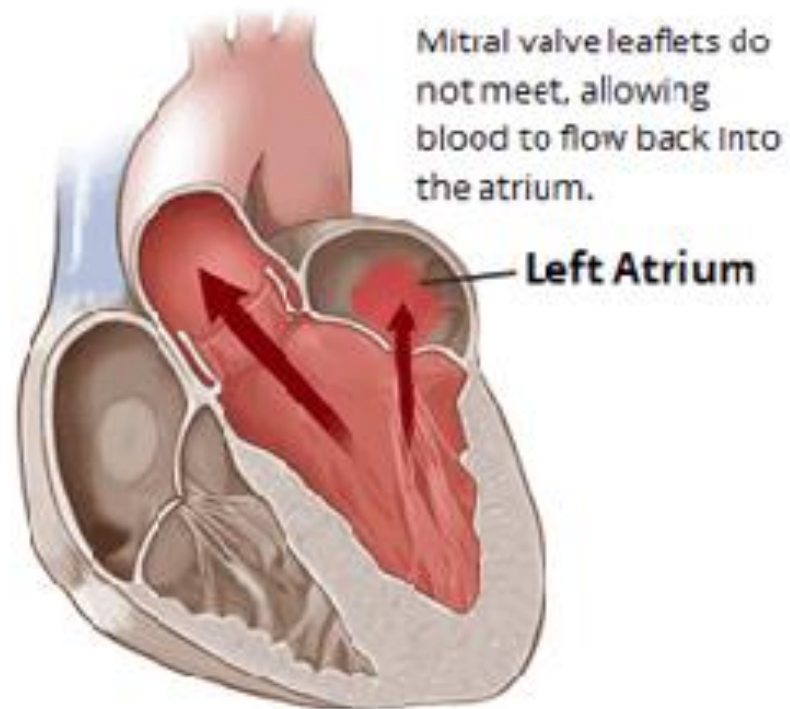
Normal Heart

Mitral Valve Stenosis

## Causes of mitral stenosis

Mitral stenosis is almost always rheumatic in origin  
There is also a rare form of congenital mitral stenosis.

## Mitral Regurgitation



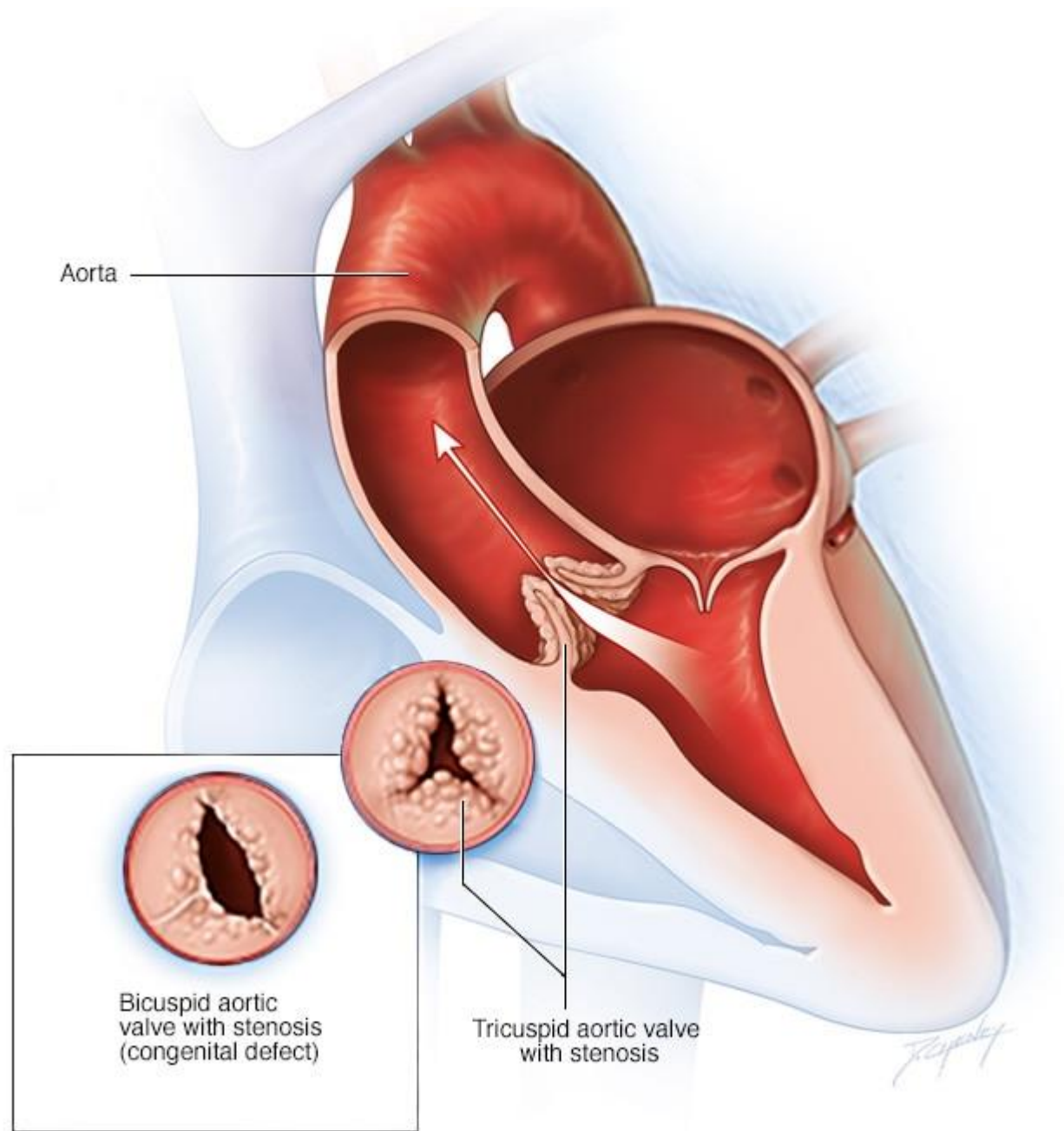
**Normal**

**Regurgitation**

**i**

## 16.80 Causes of mitral regurgitation

- Mitral valve prolapse
- Dilatation of the left ventricle and mitral valve ring (e.g. coronary artery disease, cardiomyopathy)
- Damage to valve cusps and chordae (e.g. rheumatic heart disease, endocarditis)
- Ischaemia or infarction of the papillary muscle
- Myocardial infarction



Aorta

Bicuspid aortic valve with stenosis (congenital defect)

Tricuspid aortic valve with stenosis

**i****16.84 Causes of aortic stenosis****Infants, children, adolescents**

- Congenital aortic stenosis
- Congenital subvalvular aortic stenosis
- Congenital supra- valvular aortic stenosis

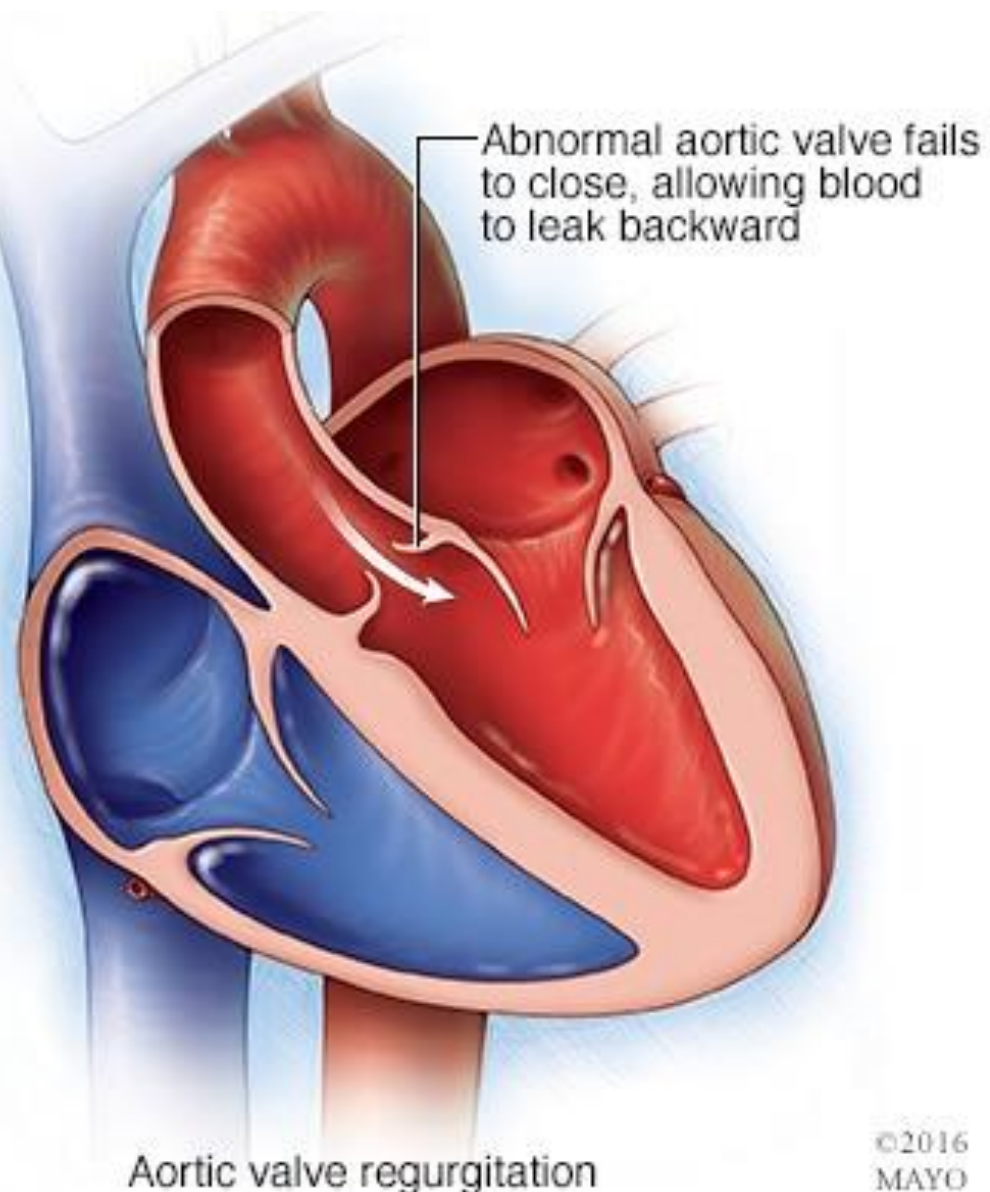
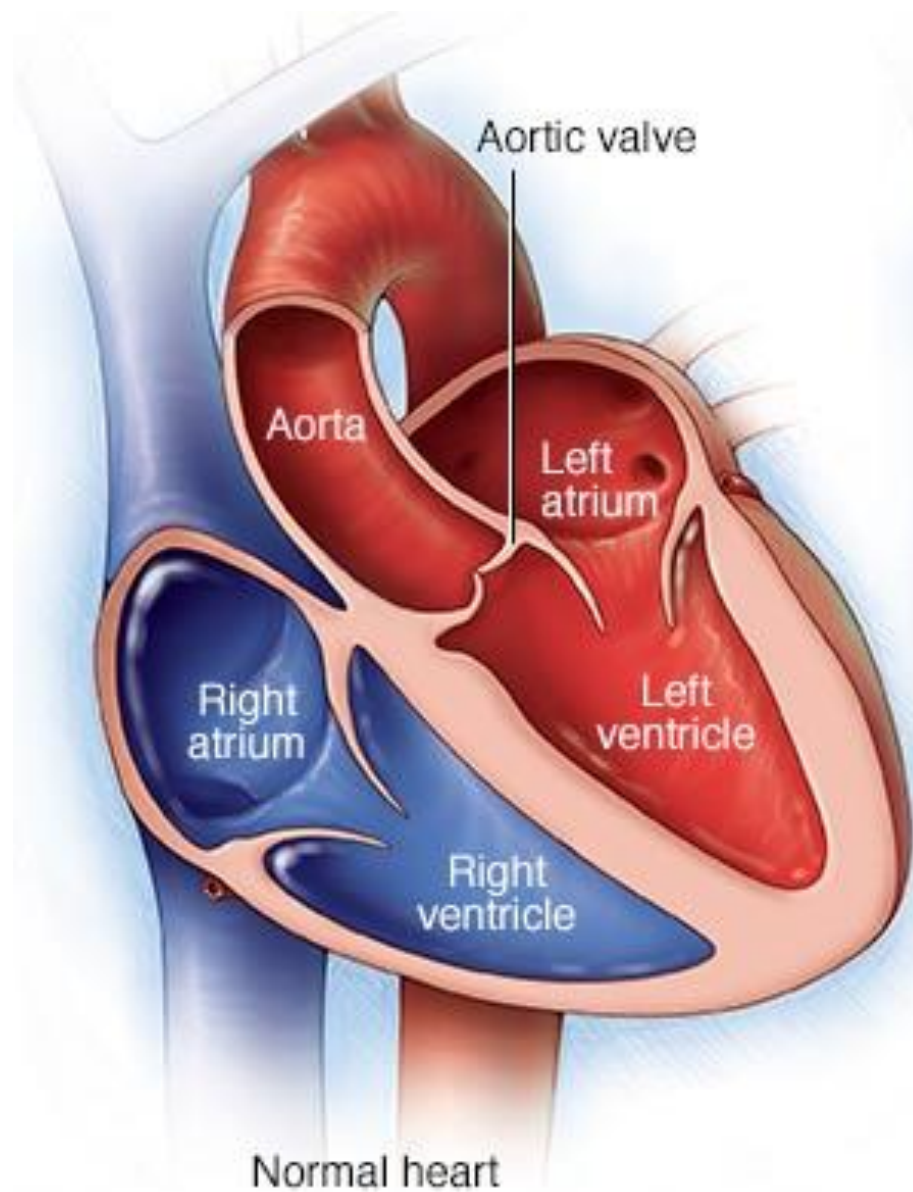
**Young adults to middle-aged**

- Calcification and fibrosis of congenitally bicuspid aortic valve
- Rheumatic aortic stenosis

**Middle-aged to elderly**

- Senile degenerative aortic stenosis
- Calcification of bicuspid valve
- Rheumatic aortic stenosis





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## 16.88 Causes of aortic regurgitation

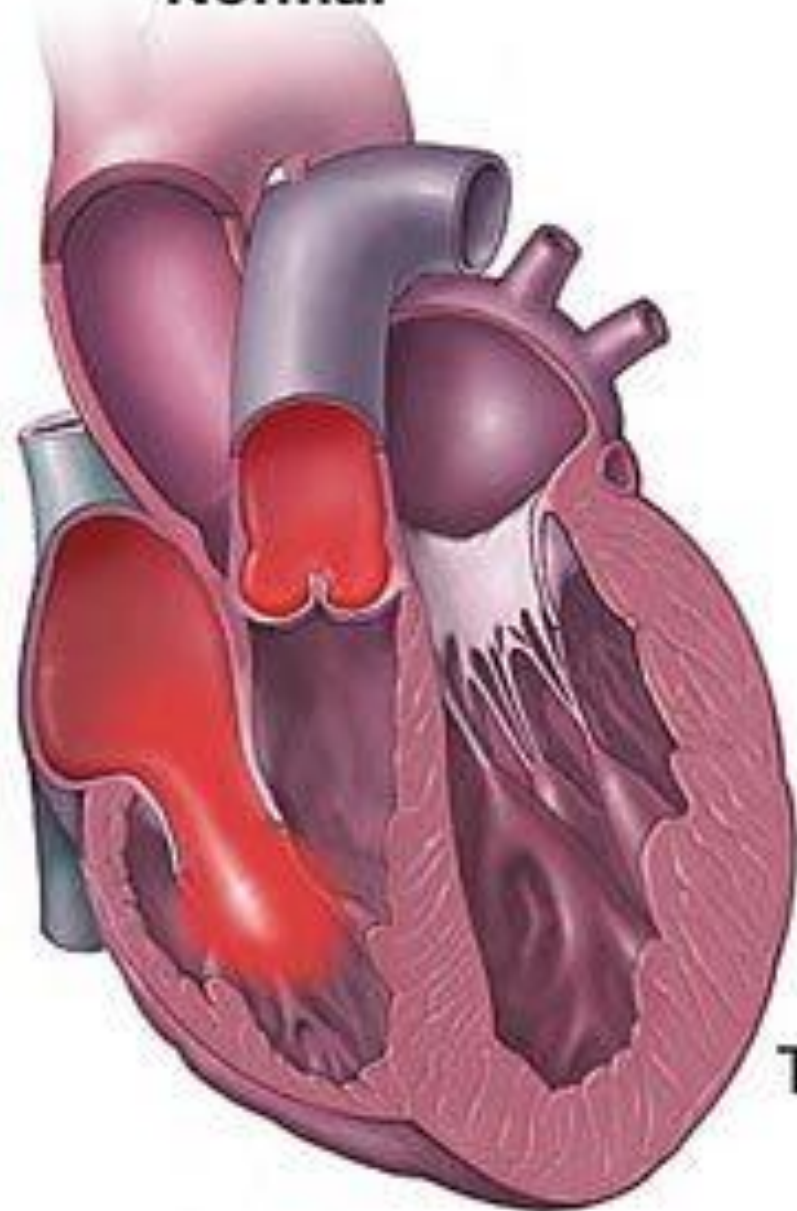
### Congenital

- Bicuspid valve or disproportionate cusps

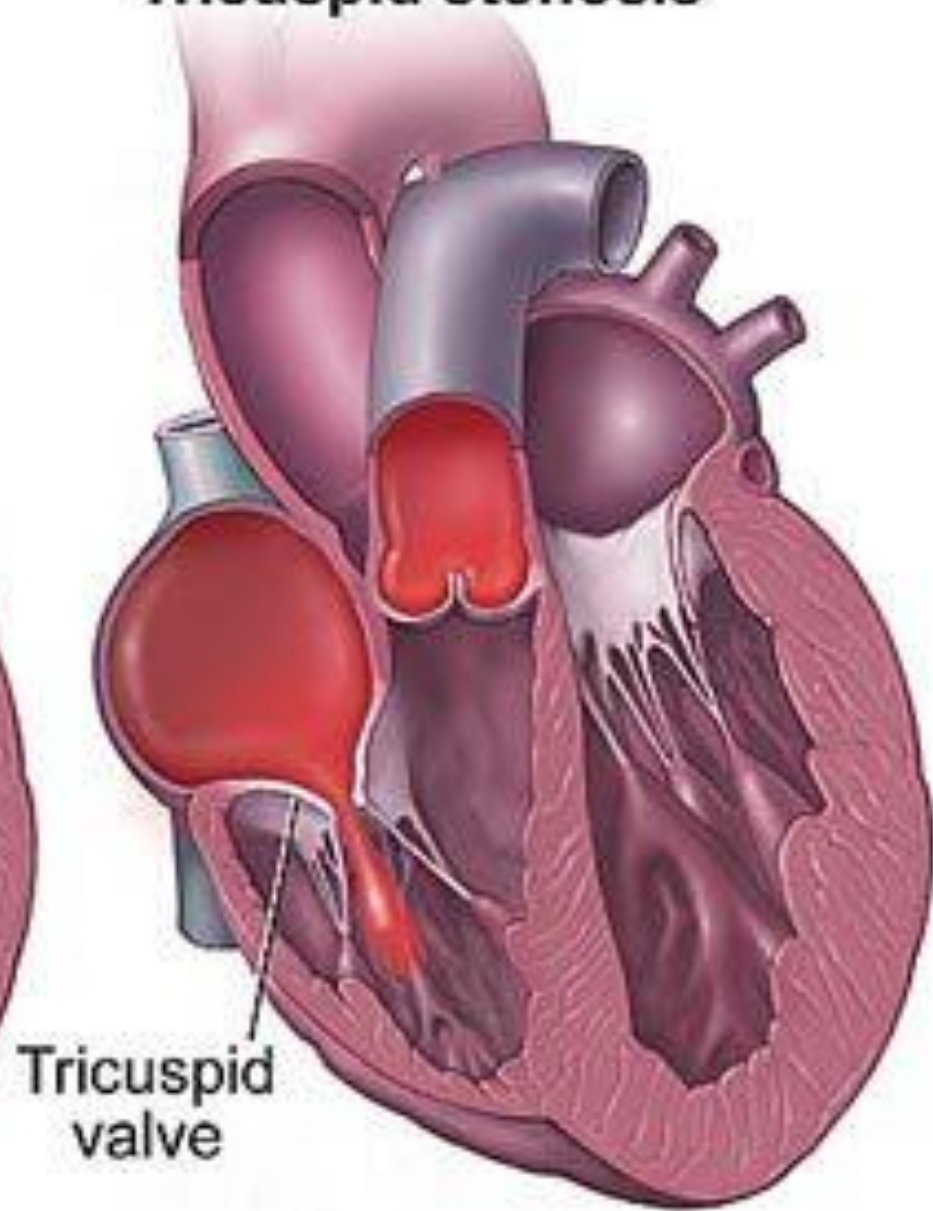
### Acquired

- Rheumatic disease
- Infective endocarditis
- Trauma
- Causes of aortic dilatation:
  - Marfan's syndrome
  - Aneurysm
  - Aortic dissection
  - Syphilis
  - Ankylosing spondylitis

**Normal**



**Tricuspid stenosis**



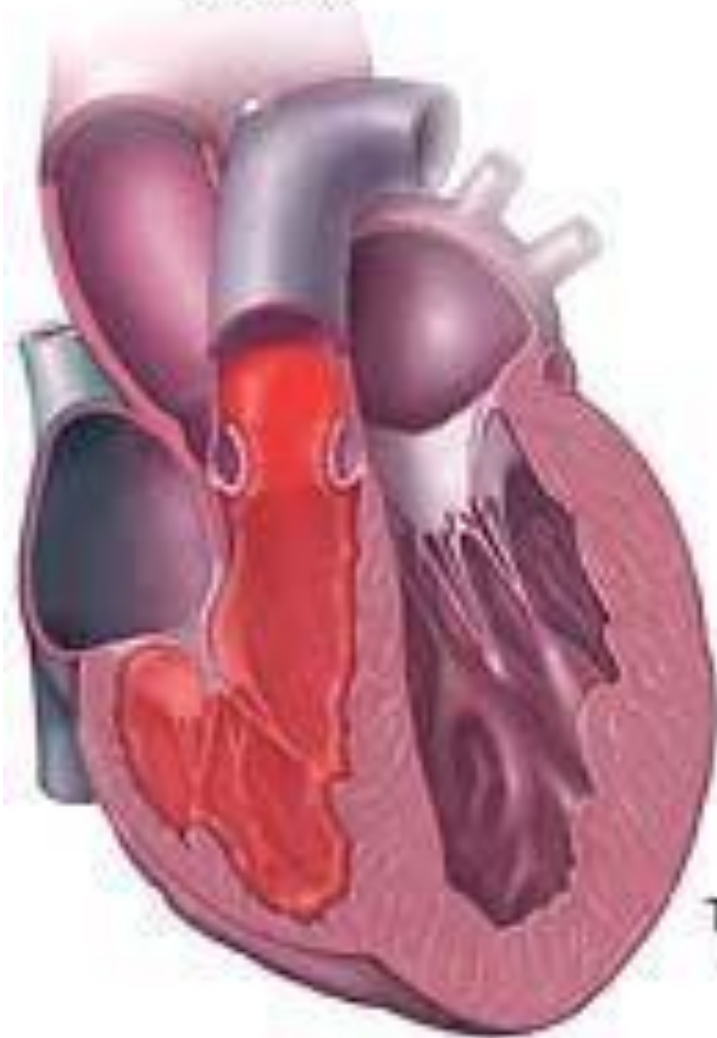
Tricuspid  
valve

## Tricuspid stenosis

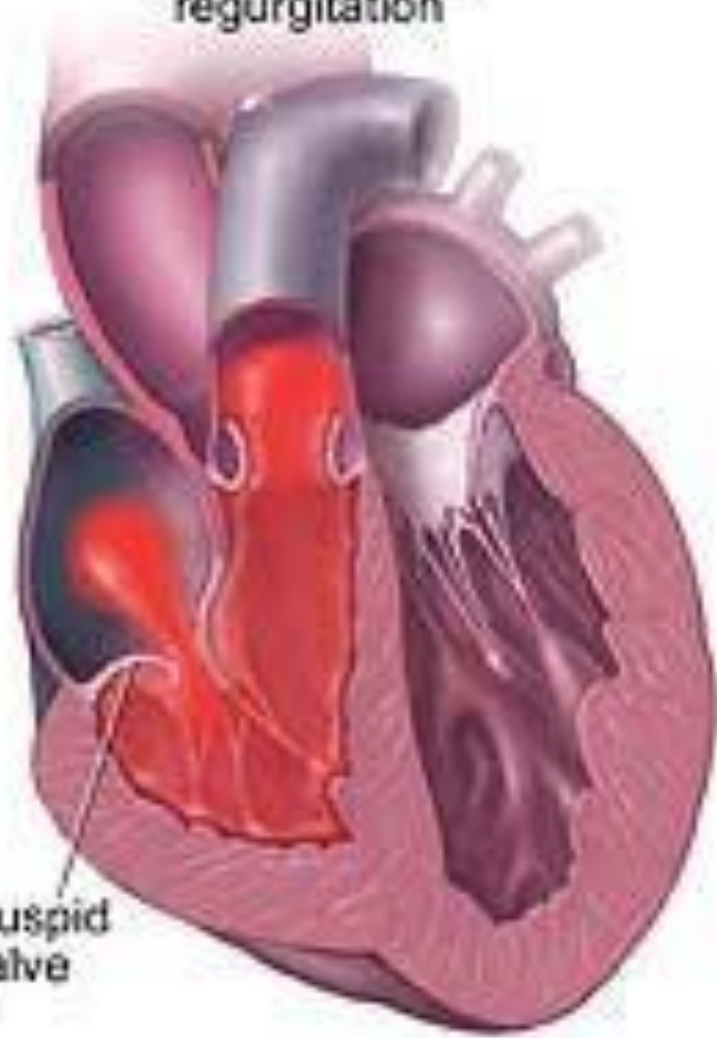
Tricuspid stenosis is usually rheumatic in origin and is rare in developed countries. It always occurs in association with mitral and aortic valve disease.

Tricuspid stenosis and regurgitation may also occur in carcinoid syndrome

Normal



Tricuspid valve regurgitation



Tricuspid valve

# i

## 16.91 Causes of tricuspid regurgitation

### Primary

- Rheumatic heart disease
- Endocarditis, particularly in intravenous drug-users
- Ebstein's congenital anomaly (see [Box 16.102](#))

### Secondary

- Right ventricular failure
- Right ventricular infarction
- Pulmonary hypertension

# Prosthetic valves

Diseased heart valves can be replaced with mechanical or biological prostheses.

All mechanical valves require long-term anticoagulation because they can cause systemic thromboembolism or may develop valve thrombosis

Biological valves have the advantage of not requiring anticoagulants to maintain proper function; however, many patients undergoing valve replacement surgery, especially mitral valve replacement, will have AF that **requires anticoagulation anyway.**

Biological valves are less durable than mechanical valves and may degenerate 7 or more years

after implantation, particularly when used in the mitral position.

They are more durable in the aortic position and in older patients, so are particularly appropriate for patients over 65 undergoing aortic valve replacement.

## Prosthetic Heart Valves



### Biologic

- Lasts 8-10 years
- No anticoagulation
- No Click



### Mechanical

- Lasts > 20 years
- Lifelong anticoagulation
- Click