Valvular heart diseases

METHODIC MATERIALS FOR INTERNATIONAL STUDENTS (IV-VI year)

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Valvular heart disease with mitral valve affection

Normal mitral valve

Anatomy

The normal mitral valve is a complex structure, consisting of leaflets, annulus, chordae tendineae, and papillary muscles. Its anatomy as studied at autopsy shows an unusual degree of variation between normal subjects. Of the two leaflets, the anterior one is the larger, both from base to margin, and also in its perimeter. It is attached to the root of the aorta and the membranous septum at the base of the heart, and is continuous with the chordae peripherally. It thus passes across the centre of the left ventricular cavity, dividing the inlet from the outlet portion. The posterior cusp is attached to the mitral ring and to the anterior cusp at both commissures. It is continuous with the posterior wall of the left atrium, and is divided into three portions by two scallops. The chordae arise from the ventricular margins of both cusps, and are inserted into the heads of the papillary muscles. There are multiple subdivisions in the chordae as they pass from papillary muscles to the cusps, which form an effective secondary pathway, additional to the main one between the cusps, for blood to enter the ventricle. There are two papillary muscles, one anteromedial and the other posterolateral. In general, the former is larger, and has a more uniform structure than the latter which may be double. Both may have up to six heads, giving rise to chordae. The papillary muscles are continuous with the trabecular and subendocardial layer of the ventricular wall. Both are supplied by a single end-artery. The mitral ring is an insubstantial structure whose function is to support valve cusps only. It is incomplete in the region of aortic root and the membranous septum. However, it is surrounded by a well developed circumferential ring of myocardium which supports it, and whose contraction during systole has the effect of significantly reducing the diameter of the valve orifice. Histologically, the normal valve cusp has a dense collagenous core, continuous with the valve ring, the valve fibrosa. This is covered on atrial and ventricular surfaces by a thin layer of loose connective tissue, and finally by endocardium.

Physiology

The normal mitral valve has a cross-sectional area of approximately 5cm2;. This allows ventricular filling to occur at a peak rate of 500 to 1000ml/s, with only a very small pressure drop across it. Left ventricular filling occurs mainly during early diastole, the rapid filling phase, and left atrial systole. During middiastole (diastasis), ventricular volume remains virtually constant, and the mitral valve itself almost closes. As heart rate increases during exercise, diastasis becomes shorter, while the duration of the rapid filling period remains virtually unchanged. At rest, approximately two-thirds of the stroke volume enters during early diastole, and the remaining one-third during left atrial systole. At rapid hearts rates, occurring at peak exercise, left ventricular filling time in normal subjects may fall to less than 100ms. If stroke volume is taken as 100ml, mean filling rate is of the order of 1 l/s, which is achieved with only a very small pressure difference between left atrium and left ventricle. This gives some indication of the effectiveness of normal mechanisms underlying left ventricular filling.

Mitral stenosis:

Definition: mitral valve stenosis is the valvular heart disease, characterized by obstruction of the blood flow form left atrium to left ventricle at the level of mitral valve.

Picture 1: mitral stenosis: view

Epidemiology

- pure stenosis: 25% of all valvular heart diseases

- mixed mitral valve diseases – 40%
- women: men ratio is 4:1

Aethiology

- chronic rheumatic heart disease
- congenital mitral stenosis may be associated with hypoplasia of the left ventricular cavity and the aorta, and also with endocardial fibroelastosis.
- **left atrial myxoma: simulates clinical picture of mitral stenosis**

In occasional patients:

- calcified mitral valve ring
- infective endocarditis, when bulky vegetations may cause obstruction to flow
- when granulomatous infiltration has occurred in association with eosinophilia.
- in nodular rheumatoid arthritis, thickening of the valve cusps has been observed, but true mitral stenosis does not result.
- in systemic lupus erythematosus, treatment of Libman-Sachs endocarditis with steroids may lead to fibrosis of the cusps with commisural fusion.
- the combination of ostium secundum atrial septal defect and mitral stenosis, Lutembacher's syndrome, is probably fortuitous.

Morphology

The stenosis may be either at valve level or below it, due to fusion of the chordae.

3 morphological variants of mitral stenosis exist:

1. Comissural: fusion of comissures; in these cases pure stenosis is usually formed

2. Valvular: the cusps changes dominate: cusps are thickened, with signs of fibrosis; may be calcified, with thrombi at atrial surfaces; usually mixed mitral valve disease is present

3. Chordal: thickening, fusion and contraction of the chordae tendineae. Chordae are shortened and fused (with each other and valve apparatus), so valve forms a rigid crater, with apex directed to left ventricle; its edges are fused and dense. With disease progression, the cusps become immobilized in this position, so not only opening, but also closing is defective and pure stenosis is changed by mixed disease: stenosis and insufficiency.

Sometimes all three forms coexist in one patient and no one is dominating.

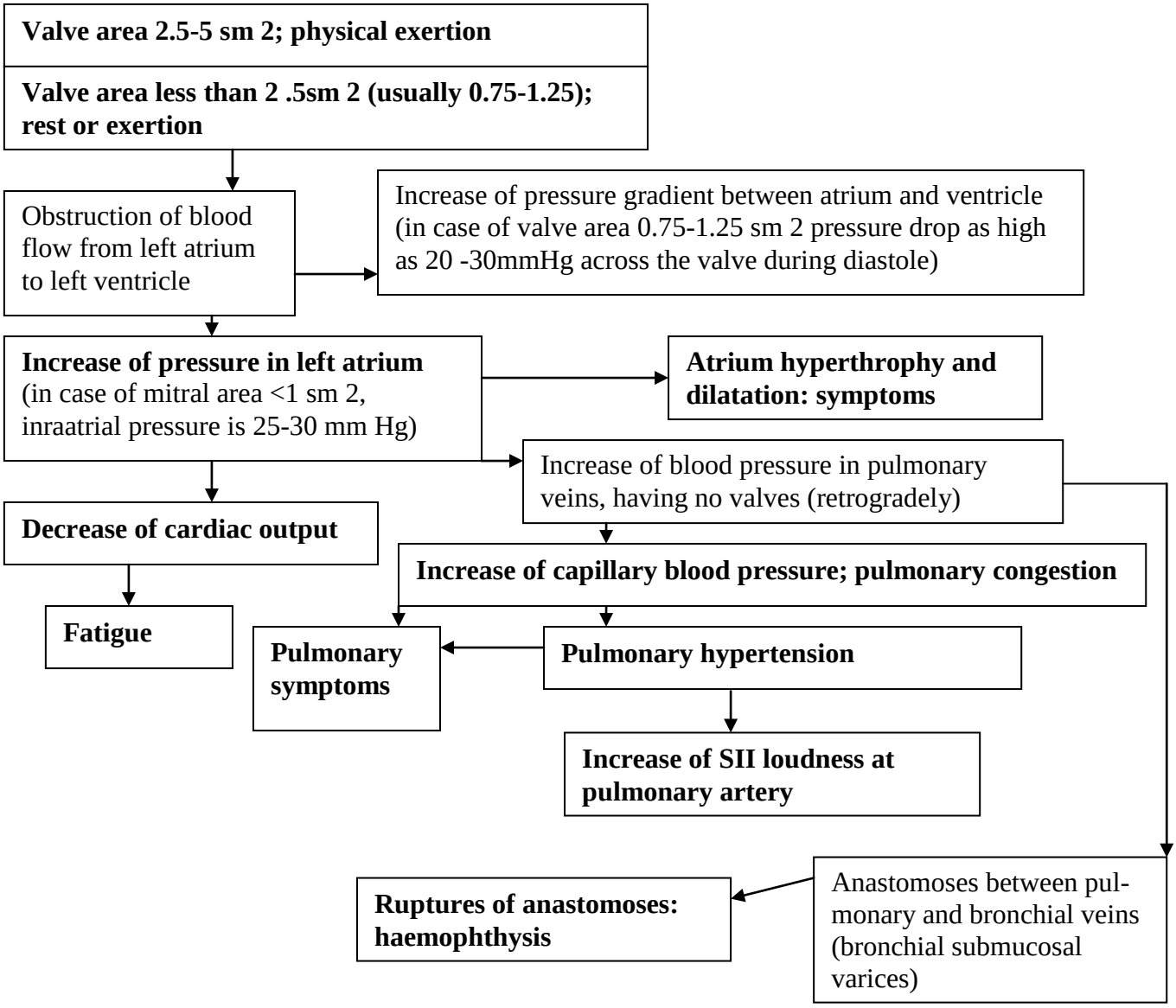
The left ventricle is usually normal or small in pure mitral stenosis, but occasionally is greatly dilated. The left atrium is characteristically enlarged: its wall may be histologically normal, but sometimes, muscle fibres are disrupted. Mural thrombosis may be present, most commonly on the free wall just above the posterior mitral valve cusp (McCallum's patch). In long-standing cases, calcification of the left atrial wall may develop in plaques on its endocardial surface. Changes of pulmonary venous congestion, pulmonary hypertension, and haemosiderosis may develop in the lungs, with dilation and hypertrophy of the right ventricle and functional tricuspid regurgitation.

Pathogenesis and clinical signs:

The normal mitral valve has a cross-sectional area of approximately 5cm2; at rest, blood flow takes place only in central part of this area (2 sm 2; pressure gradient 5 mm Hg); while in

physical exertion whole mitral valve are is used. So, if mitral valve area is 2.5-5 sm 2, no clinical signs appear at rest.

I. Early symptoms pathogenesis:



Thus, the most early symptoms, appearing in mitral stenosis, are:

1. Pulmonary symptoms:

- **Dyspnea,** at first time appearing in severe physical exertion (mitral valve area 2.5-5 sm 2); then becoming constant (further progression of valve affection, decrease of are size). Dyspnea aggravates in physical exertion and recumbent position.
- **Cough** (pulmonary congestion, pathogenesis – see heart failure)
- **Frequent respiratory infections (RVI, bronchites, pneumonias)**
- **Haemophthysis** – due to rupture of anastomoses between pulmonary and bronchial veins (due to pulmonary hypertension)

- **Night asthma-like dyspnea paroxysms, pulmonary oedema**

2. Fatigue

3. Objective symptoms:

A Left atrium enlargement:

- may be mild displacement of the left border of relative cardiac dullness to the left (in III intercostals space) **– mitral heart configuration**

B Signs of valves affection:

- **S_1 is loud and snapping** (more loud at the early stage of the diseases, is present if PQ interval is not prolonged) due to the thickening of the valve cusps and increase of pressure gradient between atrium and ventricle; decrease of blood filling in left ventricle.
- **Mitral opening snap:** sharp, high-pitched sound, heard **at the beginning of the diastole**; better heart at the lower left sternal border, at the apex, at Botkin-Erb point and radiates well to the base of the heart. Is due to vibrating of the adhered valve leaflets when opening.
- **Mesodiastolic or/and presystolic murmur** ("apical crescendo rumble") with maximum at apex, grade 1 to 4; begins after mitral opening snap separated from it by an appreciable interval. It is low-pitched and persists for a variable period throughout diastole. Murmur is caused by the turbulent blood flow from left atrium to left ventricle; **presystolic increase of murmur intensity** is present due to the increase of the blood flow while left atrium is contracting. Intensity of the murmur increases in case of increase of blood flow: physical exertion, lying position at the left side

  If the stenosis is mild, the murmur is short, but if the murmur lasts throughout diastole at a normal ventricular rate, then the degree of stenosis is likely to be at least moderately severe
- **Palpative equivalents:**

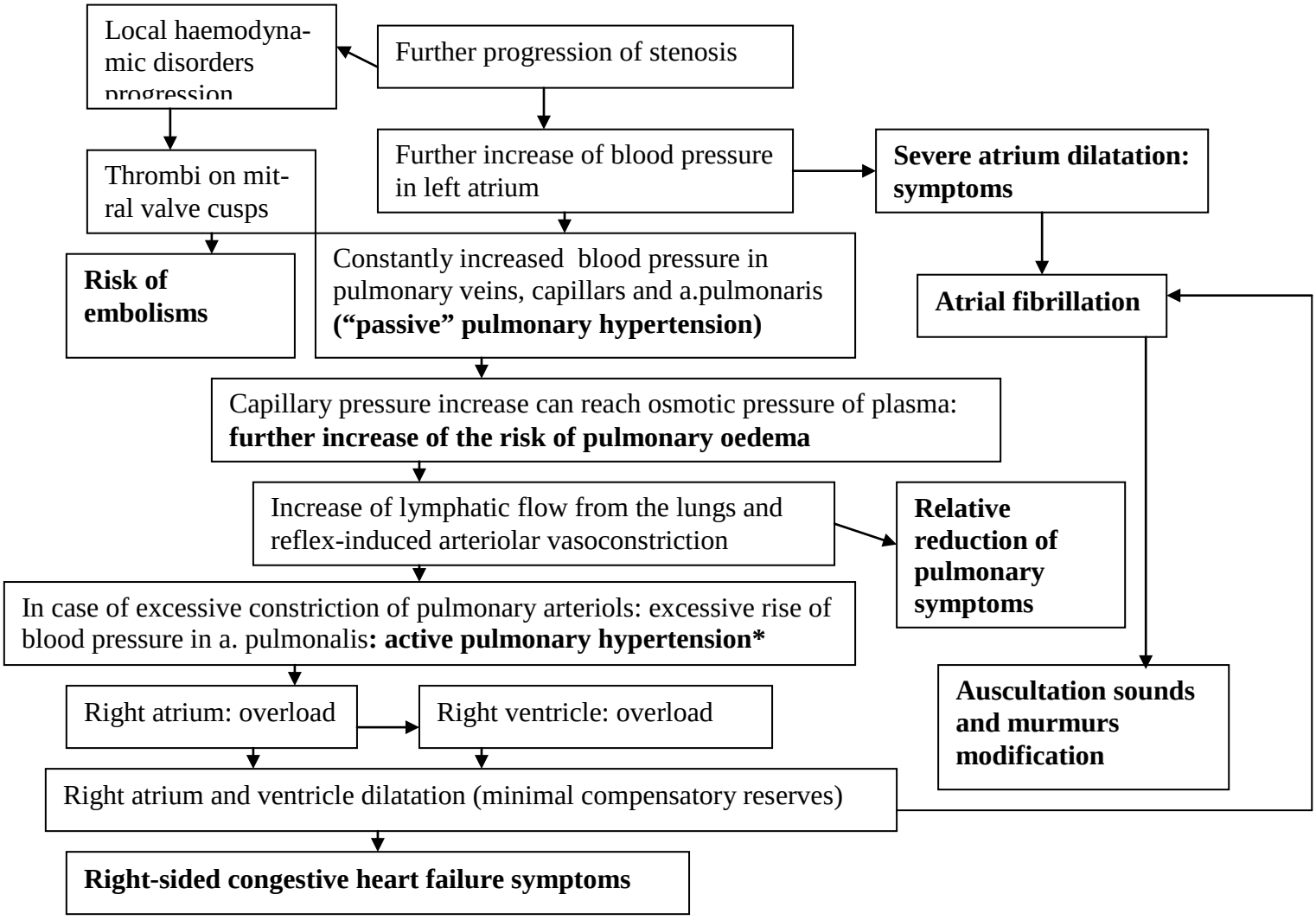
*** of the diastolic murmur: diastolic thrill at the apex** ("the cat's purr").

* **palpable SI at the apex** ("tapping apex") - typical

* **palpable opening snap -** less frequently

C. Increase of SII pulmonary component may be present

II. Further disease progression. Passive and active pulmonary hypertension. Atrial fibrillation.



*** Excessive arteriols vasoconstriction and active pulmonary hypertension are present only in some patients.**

Symptoms:

1. **Atrial fibrillation/flutter** with subsequent complains, objective and instrumental findings (50-80% of patients); before atrial fibrillation, other atrial rhythm disorders may be present: APC, paroxysmal tachycardia
2. **Right-sided congestive heart failure** with subsequent complains, objective and instrumental findings
3. **Marked left atrium dilatation** (mitral heart configuration)

4. **Right atrium and ventricle dilatation:**

- displacement of right border of relative cardiac dullness to the right
- heart push
- epigastric pulsation
- distention and pulsation of the neck veins
- relative tricuspidal regurgitation murmur: systolic murmur at the places of tricuspid valve auscultation (4th intercostal space to the right of the sternum), radiation to xiphoid area or midclavicular line, never to axilla. Variable intensity, medium pitch, blowing quality, gets louder with inspiration.

5. **Increase of a.pulmonary pressure:**

- Increase of SII pulmonary component
- pulmonary systolic ejection click: in case of severe pulmonary hypertension and marked dilatation of a.pulmonalis
- systolic murmur over a.pulmonalis (severe hypertension)
- diastolic murmur of pulmonary regurgitation: early or mid-diastolic, high-pitched, decrescendo blowing murmur increases on inspiration (Graham-Still murmur), along the left sternal border

6. **Changes in auscultation picture with progression of mitral valve affection:**

A. due to progression of valve affection

- decrease of S1 loudness (up to normal intensity): due to rigidity and fusion of the valve cusps (decrease of the amplitude of the cusp movements)
- Mitral opening snap: decrease of intensity following severe valve cusps fusion (decrease of the amplitude of the cusp movements due to fibrosis and calcification)

B. due to atrial fibrillation

- Increased loudness of S1: remaining at atrial fibrillation
- Mitral valve snap: remaining at atrial fibrillation
- presystolic increase of diastolic murmur intensity; disappears due to disappearing of active atrial systole.
- in case of rapid rhythm diastolic murmur may disappear

7. **Embolisms:** disturbances of local haemodynamic (turbulent blood flow, blood retention in auriculum of left atrium) and active pathologic process (rheumatic heart disease exacerbations), predispose to thrombi formation. Thus, recurrent pulmonary embolisms with infarctions are often present.
8. **Presence of valves affection** in case of transient bacteriemia (teeth extraction, urological and gynecological manipulations and operations etc) may **predispose to secondary**

infective endocarditis development (however, it's rather rare in pure stenosis, more frequent – in mixed valve disease).

9. **In patients, developing active pulmonary hypertension respiratory symptoms may be significantly less marked.**

Classifications of mitral stenosis:

Classification principle		
Clinical	Pressure in a.pulmonalis, mm Hg	Mitral valve area
Moderate – asymptomatic	Less than 15-19	>2 sm 2
Severe - symptoms	20-30	1-2 sm 2
Critical – symptoms, pulmonary hypertension	>30	< 1 sm 2
Complicated	-	-

Laboratory and instrumental methods in mitral stenosis diagnosis

I. ECG:

1. **left atrium hypertrophy/overload: P-mitrale**

- broad notched P waves in lead II
- inverted or biphasic P (with the inverted portion of biphasic P broader and deeper than upright portion) in V1.
- P duration is usually ≥0.12 s (normal: upper limit is 0.11)

2. **right atrium hypertrophy/overload: P-pulmonale**

- tall peaked P waves (≥0.25mV) most prominent in II and V1

3. **right ventricle hypertrophy/overload:**

- tall R waves in V1 (≥0.5mV); in less extreme degrees – moderately deep S wave in V1 with R wave voltage exceeding S wave voltage;
- abnormal S wave in V5 or V6 (≥0.7mV); in less extreme degrees – prominent terminal S waves in V5 or V6
- shift of the mean QRS axis to the right

4. **rhythm disorders**

II. **X-ray**

- increase of left atrium size

Usually: enlargement of the left atrium that is selective, i.e. proportionately greater than that of the heart shadow as a whole. This appears on the penetrated posteroanterior film as a double outline on the right side of the heart shadow, with elevation of the left main bronchus, and enlargement of the left atrial appendix which forms that part of the left heart border just below the main pulmonary artery.

- Mitral valve calcification: may be visible on the posteroanterior film just to the left of the spine, on the continuation of the shadow of the left atrium.
- in severe cases – increase of right ventricle and a.pulmonalis size
- interstitial pulmonary oedema signs (Kerly lines)

III. **Phonocardiography:** detailed sounds picture, confirming auscultation results

IV. **Echocardiography:**

- area of mitral valve square
- gradient of pressure between left atrium and ventricle
- transmitral blood flow
- blood pressure in a.pulmonale
- severity of leaflets calcification, deformity and movement disorders: The valve is thickened, opens poorly, and closes slowly. The anterior and posterior leaflets are fixed and move together, rather than in opposite directions.
- grade of chordae shortening
- size of left atrium, presence of thrombi
- sizes of right atrium, left and right ventricles
- severity of local haemodynamic disorders (presence of regurgitations, including relative ones)

Description of picture (also see picture 1):"Doming" of the leaflet opening during diastole in seen at the long axis view. Similarly, in the 4 chamber apical view, the leaflets arch into a dome-shape during diastole. The Color Doppler Imaging shows a funneling of the diastolic flow through the narrowed orifice. M-mode diastolic patterns show the prolonged diastolic E-F slope. The normal mitral valve has a valve area in excess of 4 cm sq. Symptomatic mitral stenosis readily occurs when the valve area falls below 1.4 cm sq

Picture 4 (a,b,c):

4.a Dimensional parasternal long-axis view of a patient with mitral stenosis, showing thickened valve cusps (white arrow), with poor leaflet separation in diastole. Left atrium is enlarged, with a thrombus in the posterior aspect of it (black arrow). Aortic valve is also stenotic

4.b. Diastolic color Doppler flow convergence in the apical four chamber view of mitral stenosis.

4.c. *Hockey stick* appearance of the anterior mitral leaflet in mitral stenosis (white arrow)

Due to highly informative results of echocardiography, no invasive methods should be used to confirm the findings.

Diagnosis formula:

Rheumatic heart disease, active phase. Mitral stenosis. Atrial fibrillation. Congestive heart failure II A stage; III functional class.

Differential diagnosis:

- Myxoma of the left atrium: auscultation results change during changing of patient's position

	Myxoma (75% - left atrium, prolapse to left ventricle may occur)	Mitral stenosis
Complains and history	- fever, fatigue, arthralgias, pallor - increasing dyspnoea - systemic embolization (thrombi on myxoma surface)	- intoxication may be due to activity of rheumatic heart disease
Ausculta-tion	- delayed diastolic and presystolic murmurs - loud first sound, - opening snap(tumour plop) - systolic murmur of mitral regurgitation may be present (valve disfunction) **!!! auscultation results change with changing of the position of patient**	Same **Auscultation results don't depend on patient's position**
	- erythrocyte sedimentation rate and plasma proteins are frequently, but not invariably, abnormal.	Same in active rheumatic heart disease
Echocardiography confirms the diagnosis		

- **Atrial septum defect**: wide fixed splitting of SII, ejection click and murmur at a.pulmonale, diastolic overload of the right ventricle
- **Cor triatriatum:** a rare cause of obstruction to blood flow at left atrial level and may mimic mitral stenosis, particularly in childhood.
- **Pulmonary veno-occlusive disease:** may also present as silent mitral stenosis, often with a raised pulmonary wedge but a normal left atrial pressure.

Final diagnosis: echocardiography

Natural course:

Progression of the valve affection. Atrial fibrillation or pregnancy (associated increase in cardiac output and the transmitral pressure gradient often precipitates symptoms) lead to more severe symptoms progression. More quick progression is observed in patients with severe deformation of cusps and subvalvular structures.

Complications: arrhythmias (typical – atrial fibrillation/flutter); embolisms, pulmonary oedema, predisposition to pulmonary inflammatory diseases; infective endocarditis.

Prognosis: in some patients with mild and moderate stenosis without progression the long-time asymptomatic course may occur (up to 20 years); but in general prognosis is unfavorable without timely surgical correction.

Treatment: surgical

Indications to surgical treatment:

- first decompensation symptoms in patients with mitral valve area <1sm 2
- asymptomatic severe stenosis with severe pulmonary hypertension >60 mm Hg

Surgical treatment:

Open mitral commissurotomy may be effective in patients without substantial mitral regurgitation.

Percutaneous transvenous mitral commissurotomy (PTMC) with Inoue balloon catheter

Replacement of the valve is indicated when combined stenosis and insufficiency are present or when the mitral valve is so distorted and calcified that a satisfactory valvulotomy is not possible. Operative mortality rates are low: 1–3% in most institutions.

Problems associated with prosthetic valves are thrombosis (especially at the mitral position), paravalvular leak, endocarditis, and degenerative changes in tissue valves. Warfarin anticoagulant therapy is mandatory with mechanical prostheses and is usually employed for at least the initial 3 months with bioprostheses, especially if the patient has significant left atrial enlargement. If atrial fibrillation persists postoperatively, ongoing anticoagulation is required.

Balloon valvuloplasty is becoming increasingly popular in patients without accompanying regurgitation.

However, after successful surgical treatment restenosis is possible, but its rate is lower than that with aortic stenosis.

If surgical treatment is impossible by any reason, treatment of congestive heart failure, arrhythmias and embolisms is necessary.

Mitral regurgitation (Mitral Insufficiency)

Definition: mitral regurgitation (mitral insufficiency) is a vlave disease with structural changes of the mitral valve as a whole or its separate parts, causing blood regurgitation from left ventricle to left atrium, appearing at every systole.

Prevalence:

Including the rate of mitral prolapse – 2-10%. Mitral prolapse more often occurs in children and youths; men: women ratio is 0.3. In adults and aged with mitral regurgitation men: women ratio is about 1:1.

Aethiology:

- rheumatic disease (50%) associated with a thickened valve with reduced mobility and often a mixed picture of stenosis and regurgitation (90%)

- myxomatous degeneration (eg, mitral valve prolapse with or without connective tissue diseases such as Marfan's syndrome, pseudoxanthoma elasticum, Ehlers-Danos syndrome, and osteogenesis imperfecta.)
- infective endocarditis
- subvalvular dysfunction (due to papillary muscle dysfunction or ruptured chordae tendineae)
- severe left ventricle dilatation
- cardiac tumors, chiefly left atrial myxoma, are a rare cause of mitral regurgitation.
- mitral valve ring calcinosis
- congenital splitting of mitral valve cusp, usually associated with interatrial septum defect

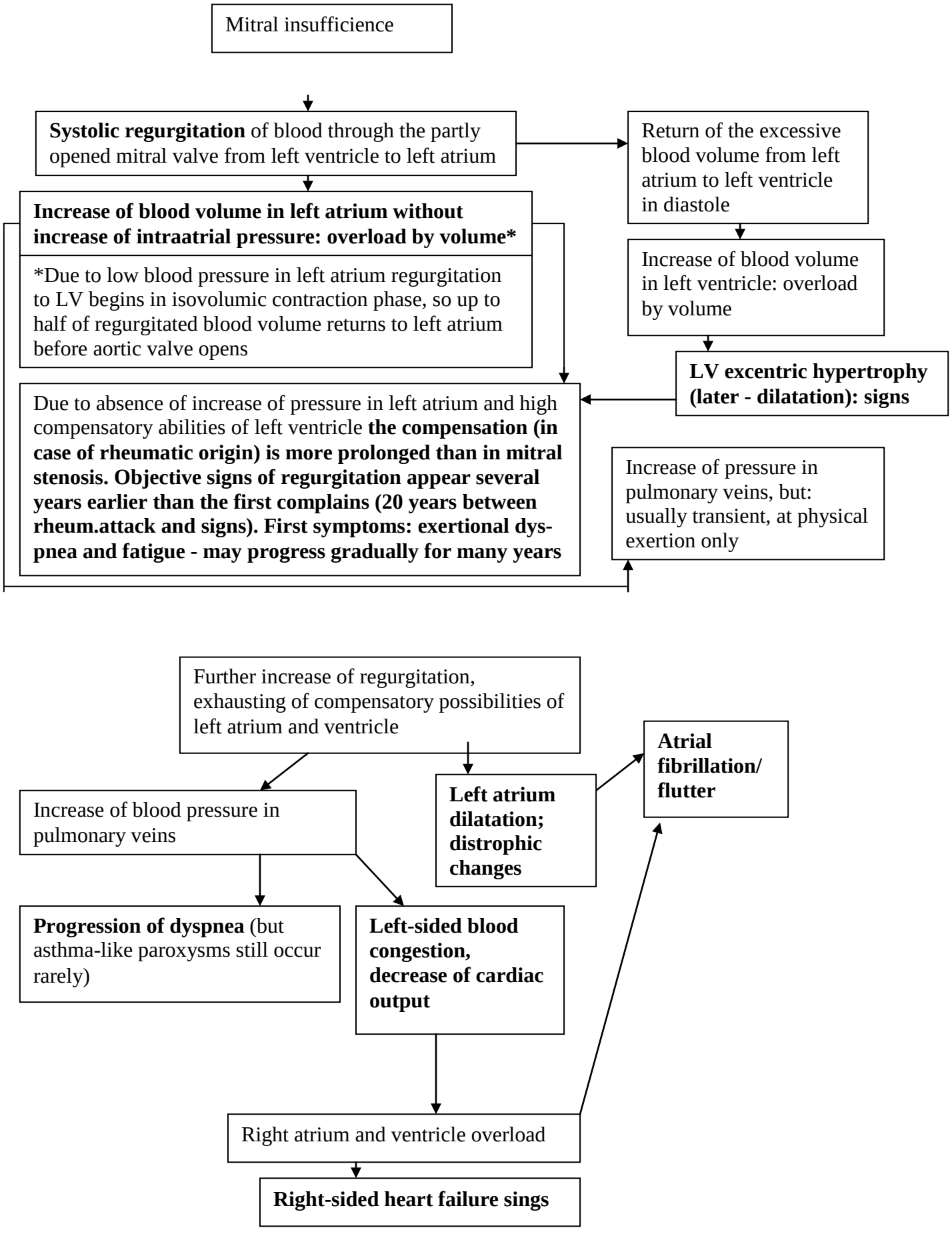
Morphology:

Rheumatic heart disease: thickened, deformed, shortened cusps with signs of calcinosis at their base.

Mitral valve prolapse (floppy mitral valve): non-inflammatory process which may affect either cusp, partially or completely.

- Increase in cusp area, causing folding and upward doming into the left atrium during systole.
- The chordae may become elongated, tortuous, and thinned, predisposing to chordal rupture.
- The abnormal chordae can undergo fibrosis, as can the cusps, leading to an erroneous diagnosis of chronic rheumatic involvement.
- Ulceration of the cusps may also occur, predisposing to thrombosis on their surface, and also to infective endocarditis.
- The ring circumference may be normal or increased.
- The papillary muscles are normal.
- Histologically, the central valve fibrosa is abnormal with large areas in which fibrous tissue is either absent altogether, or where the collagen bundles are fragmented, coiled, or disrupted. These lie in pools of abnormal acid mucopolysaccharide. A dense layer of laminated collagen forms over the atrial surface of the cusp.
- There is no evidence of vascularization or of inflammatory cells in the absence of secondary infective endocarditis.

Pathogenesis and clinical symptoms:



The time of the clinical symptoms appearance, acuteness and severity of clinical symptoms depend on the cause of mitral regurgitation.

In cases of rheumatic heart disease:

1. Late symptoms appearance: time between rheumatic heart disease/rheumatic fever attack usually exceeds 20 years
2. Usually objective signs of valve affection exceed appearance of complains (time period – several years)
3. Progression of symptoms is rather slow
4. All these features are due to absence of increase of intraatrial blood pressure and high compensatory possibilities of the left ventricle

In case of acute mitral insufficience (rupture of chordae, papillary muscle tearing off):
Acute, sudden regurgitation appearance leads to acute increase of blood pressure in left atrium thus to subsequent acute increase of blood pressure in pulmonary veins and symptoms of pulmonary oedema.

Symptoms:

1. Pulmonary symptoms:

- exertional dyspnea
- rare – asthma-like dyspnea paroxysms

2. **Atrial rhythm disorders (up to fibrillation/flutter)**
3. **Fatigue (due to low cardiac output)**
4. **Right-sided congestive heart failure (usually appears late)**
5. **Objective signs of valve affection:**

A. Left atrium and left ventricle enlargement:

- displacement of the left border of relative cardiac dullness to the left upwards (in III intercostals space) and to the left in V intercostals space
- displacement of the apex beat to the left

B Signs of valves affection:

- **S_1 may be weak** or absent or buried in systolic murmur
- **SII is splitted** due to quick ejection of blood from left ventricle (about half of blood volume returns to atrium) and thus more early aortic valve closing
- **SIII is present at the apex** (with palpable equivalent); 0.12-0.17s after aortic valve closure sound at the completion of the rapid-filling phase of the mitral valve, is believed

to be caused by sudden tensing of the papillary muscles, chordae tendiniae and valve leaflets.

- **Presystolic gallop rhythm at the apex**
- **Systolic regurgitation murmur:** III/IV grade of intensity or louder**,** more intensive at the apex, radiated to axillar region; pansystolic or decrescendo

- **Rheumatic valve affection:** pansistolic blowing murmur, high-pitched; with stable loudness. Murmur is heard over the wide area, radiates to axillar region and left subscapular region. Murmur intensity doesn't depend on presence of sinus rhythm or aftrial fibrillation. Increase of murmur intensity is observed at inspiration and decrease – if venous return to the heart is reduced. Moderate correlation between murmur intensity and regurgitation degree exists.
- **Rupture of chordae tendineae or primary involvement of the posterior leaflet:** radiation to the base of heart
- **Mitral prolapse:** see below

C. **Signs of pulmonary hypertension if present:**

- **Pulmonary component of SII** louder (SII accent on pulmonary artery)
- **Other signs (see mitral stenosis) may be present if hypertension is severe**

D. **Palpable equivalents:**

*** of the systolic murmur: systolic thrill at the apex** ("the cat's purr").

* **palpable SIII at the apex**

E. Embolisms may be present (especially in patients with atrial fibrillation)

F. Infective endocarditis: mitral valve insufficiency predisposed to infective valve affection

Mitral prolapse (=systolic click-murmur syndrome, Barlow's syndrome, floppy-valve syndrome, billowing mitral leaflet syndrome):

- **Most patients** are female (prevalence in healthy young women – 10%), many are thin, and some have minor chest wall deformities.
- **Complains**: nonspecific chest pain, dyspnea, fatigue, or palpitations (VPC); syncopes or presyncopes.
- **Objective findings:**

* Sizes of the heart are unchanged.

* Auscultation: present normal SI, short late high-pitched systolic murmur preceded by one or several systolic clicks (mid- or late). These findings are accentuated in the standing position, Valsava maneuver.

- in occasional patients with mitral valve prolapse (especially these with pansystolic murmur) the course may be more severe, including possibility of chordae tendinea

rupture, infective endocarditis, rhythm disorders (supraventricular and ventricular tachycardia) and embolic cerebrovascular events. About 2% of patients (more often men, more often over 60) require surgical treatment. Echocardiographic evidence of marked thickening or redundancy of the valve is associated with a higher incidence of most complications.

Some other causes of mitral regurgitation:

Papillary muscle dysfunction

In part, the position of the cusps is maintained during systole by contraction of the papillary muscles as the left ventricular cavity gets smaller. The main causes are:

- affection by ischaemic or other left ventricular disease with impaired ability to contract.
- If left ventricular cavity size is greatly increased, the relation between wall movement and papillary muscle shortening becomes abnormal.

The clinical picture, therefore, is usually dominated by the main disease of left ventricle. The presence of mitral regurgitation is demonstrated by either a late or a pansystolic murmur, which often varies in its intensity and timing from day to day, and which becomes softer with successful treatment of the underlying condition.

Ruptured papillary muscle

This is a **rare complication of acute myocardial infarction, causing a sudden deterioration in the patient's clinical condition.** Complete rupture of a papillary muscle may occur, or less commonly, only a single head may be involved. **Complete rupture** usually occurs 2 to 5days after the infarction, and is rarely associated with survival for more than 24 or 48 h without very prompt surgical intervention. Death is due to cardiogenic shock and pulmonary oedema. A pansystolic murmur may sometimes be audible at the apex. Partial rupture, i.e. loss of one of the heads, occurs rather later after the infarct and, like complete rupture, causes a striking deterioration in clinical state, along with the development of a pansystolic murmur, but in this case more prolonged survival is possible. The posteromedial papillary muscle is involved more frequently than the anterolateral, by both partial and complete rupture. When complete rupture occurs, death usually occurs before definitive treatment can be undertaken, but partial rupture can be diagnosed by cross-sectional echocardiography and potentially treated by early mitral valve replacement, once the haemodynamic situation has been stabilized. The prognosis, however, is significantly worse than that after chordal rupture due to severe left ventricular disease.

Endomyocardial fibrosis (restrictive cardyomyopathy)

This is a disease characterized by fibrosis of the endocardium and underlying myocardium of either or both ventricles. It is common in Uganda and surrounding countries in East Africa where

it accounts for approximately 10 per cent of hospital admissions with heart disease, and in Nigeria in West Africa. It also occurs, less commonly, in South India and Sri Lanka. Occasionally, it is seen in Europeans who have lived in affected areas and very rarely in those who have never been to the tropics. **In case of left ventricle changes dominating, posterior mitral valve cusp and its papillary muscle are affected.**

Mitral ring calcification

Patients: elderly, more common in females.

Aethiology: degenerative condition, it occurs more frequently when the resistance to left ventricular ejection is increased, such as with aortic stenosis or hypertension.

Symptoms: usually causes no symptoms, being detected incidentally by the presence of calcification in the mitral ring on chest radiography or on echocardiography.

Complications: Yet it is not a totally benign condition. It is a potential source of systemic emboli, and a focus for infective endocarditis. Approximately half the patients have abnormalities of conduction, including high-grade atrioventricular block, sinus node disease or bundle branch block. Mild mitral regurgitation is common, but rarely is it severe enough to need valve replacement.

Treatment: only infective endocarditis prevention

Laboratory and instrumental diagnosis:

ECG:

- left atrium overload/hypertrophy
- left ventricle overload/hypertrophy
- increase of voltage in leads, reflecting the electric activity of left ventricle
- shift of the frontal plane QRS axis to the left
- S wave in V1 or V2>2.5 mV
- R wave in V5 or V6 >2.5 mV
- Downslopping ST and asymmetrically inverted T wave

Phonocardiograhpy: confirms the auscultation results

X-ray:

- increase of left ventricle and left atrium
- in long-term disease – increase of the right chambers and pulmonary congestion

Echocardiography:

- sizes of heart chambers
- valve leaflets and chordae condition
- severity of mitral regurgitation:

- mild: < 30%
- moderate: 30-50%
- severe: >50%

- underlying pathologic process
- pulmonary artery pressure

Diagnostic formula:

1. Rheumatic heart disease, inactive. Mitral regurgitation (severe). Atrial fibrillation. Congestive heart failure II B stage (III functional class)
2. Marfan syndrome. Mitral valve prolapse (insignificant).

Differential diagnosis:

1. Differential diagnosis with functional (innocent) systolic murmurs
2. Differential diagnosis with other diseases causing systolic murmur

Echocardiography finally confirm presence of mitral regurgitation.

3. Diagnosis of the aethiology of valve affection.

Course: usually benign, especially in case of insignificant or mild mitral regurgitation. Compensation may last whole life. In severe or acute insufficiency cause of death may be congestive heart failure, pulmonary oedema, rarely embolisms.

Treatment

Surgical – main method of treatment.

Indications:

- first decompensation symptoms in patients with chronic mitral regurgitation
- acute mitral regurgitation
- even without signs of decompensation: severe mitral regurgitation – end-systolic size of left ventricle >4.0-4.5 sm, end-diastolic volume of left ventricle index > 40-50 ml/m 2
- infective endocarditis in case of antibacterial treatment inefficacy

Methods:

- prosthetic valve (mitral valve replacement)
- mitral valve repair (plastic operations)

Aortic stenosis

Definition: aortic stenosis is the consequence of a fixed obstruction to left ventricular ejection. The obstruction is most commonly at the level of the valve itself, but may also be immediately above the sinuses or within the left ventricle.

Prevalence

True prevalence of aortic stenosis is unknown

The prevalence of sclerodenerative stenosis: approximately 25% of patients over age 65 and 35% of those over age 70 have echocardiographic evidence of sclerosis, with 2–3% of these exhibiting hemodynamic evidence of stenosis. Degenerative valve disease is three to four times more frequent in men than in women and is more common in smokers and hypertensives.

Aetiology

- rheumatic – 24%
- degenerative senile with calcinosis – 33%
- fibrosis of congenitally bicuspid valve – 38%
- congenital – 2%
- atherosclerosis-induced (II type hyperlipidemia) - <2%

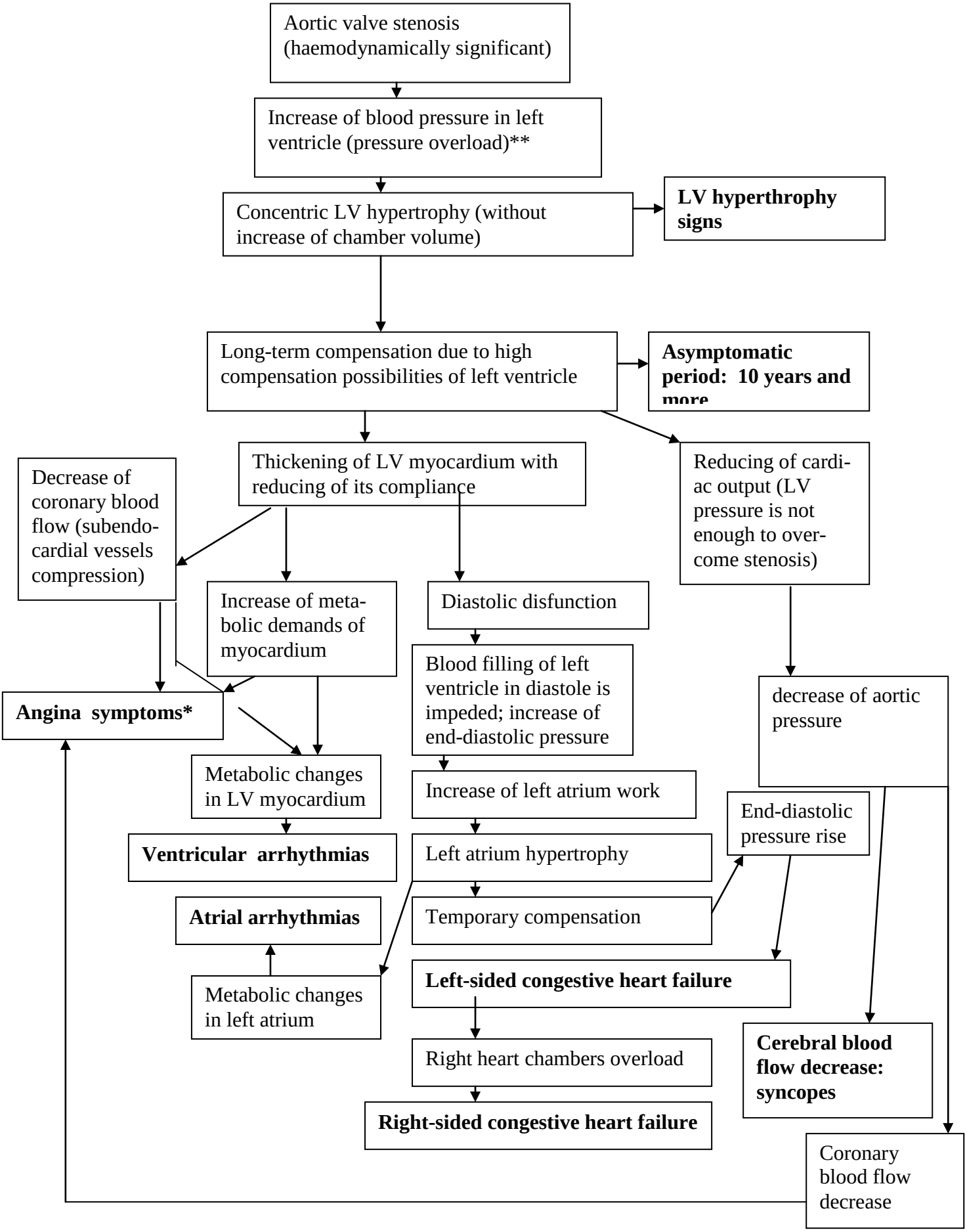
Isolated subaortic stenosis (asymmetric hyperthrophic cardiomyopathy) cause symptoms similar to these in aortic stenosis.

Morphology:

Fusion of cusps edges, cusps thickening and deformation. In calcinosis calcification of aorta ring and leaflets bases is present, and in bicuspidal valve calcinosis signs are found directly in leaflets.

Pathogenesis and clinical symptoms:

Normal aortal valve size is 3-5 sm 2; its narrowing even up to 50% doesn't cause clinical symptoms, except turbulent blood flow revealed as a murmur. Haemodynamically significant stenosis degree is revealed in case of aortic valve area size 30% of normal or less.


Aortic valve stenosis (haemodynamically significant)
Increase of blood pressure in left ventricle (pressure overload)**
Concentric LV hypertrophy (without increase of chamber volume)
LV hyperthrophy signs
Long-term compensation due to high compensation possibilities of left ventricle
Asymptomatic period: 10 years and more
Thickening of LV myocardium with reducing of its compliance
Reducing of cardi-ac output (LV pressure is not enough to over-come stenosis)
Decrease of coronary blood flow (subendo-cardial vessels compression)
Increase of meta-bolic demands of myocardium
Diastolic disfunction
Blood filling of left ventricle in diastole is impeded; increase of end-diastolic pressure
decrease of aortic pressure
Angina symptoms*
Metabolic changes in LV myocardium
Increase of left atrium work
End-diastolic pressure rise
Ventricular arrhythmias
Left atrium hypertrophy
Atrial arrhythmias
Temporary compensation
Left-sided congestive heart failure
Metabolic changes in left atrium
Cerebral blood flow decrease: syncopes
Right heart chambers overload
Right-sided congestive heart failure
Coronary blood flow decrease

* **angina symptoms** are also caused by decrease of blood flow to ostia of coronary arteries (Ventura phaenomena) accompanying severe blood flow acceleration through the narrowed aortic valve (fall of the near-wall pressure in aorta).

**systolic pressure drop between the left ventricular cavity and the aorta, which in symptomatic cases, may be greater than 50 to 70mmHg at rest, and reach over 200mmHg on exertion.

Thus, following symptoms are revealed:

1. The earliest is **dyspnea**, gradually increasing with decrease of physical exertion tolerance; paroxysmal nocturnal dyspnea also may occur at the late stages of the disease.
2. **Cardiac pain:** more often cardialgias, but also angina symptoms, indistinguishable from these in IHD patients
3. **Syncopes, dizziness, lightheadedness:**

- **at exertion:** caused by decrease of cerebral blood flow hypotension resulting from the combination of exercise-induced vasodilation and a fixed cardiac output.
- **at rest:** caused by ventricular arrhythmias (short periods of ventricular tachycardia) or transient complete atrioventricular block due to involvement of the atrioventricular node by calcification
- **Arrthythmias:** more early – ventricular, later (atrial overload) – supraventricular

In the majority of cases, calcification is confined to the aortic valve, but in a minority it may spread to involve the anterior cusp of the mitral valve or the atrioventricular node, and thus give rise to a prolonged P-R interval or even to complete heart block.

4. **Objective symptoms:**

A. Systemic haemodynamic changes

- decrease of systolic blood pressure
- small pulse (pulsus tardus et parvus)
- The carotid pulse is slow rising with a reduced amplitude and an early notch on the upstroke, followed by a thrill.

B. Heart chambers dilatation

- displacement of left border of relative cardiac dullness (to the left and downwards)
- apex beat is active, heaving and displaced to the left and downwards

C. Valves affection

- SI is normal or soft
- A_2 is increased (atherosclerosis) or diminished (rheumatism); splitting of SII may be present in young patients with mobile cusps (delayed aortic valve closure tone).
- Systolic (crescendo-decrescendo) murmur in the 2^{nd} right and 3^{rd} left intercostals space, radiation to neck and down left sternal border, often loud and with a thrill, medium pitch,

harsh quality, heard best with sitting and leaning forward. **Further reduction of the stroke volume leads to decrease of murmur intensity.**

- Aortic ejection click may be heard just before the murmur
- III tone may be present; in congestive heart failure – IV tone.
- An additional short, soft early diastolic murmur is nearly always present, although this does not imply haemodynamically significant aortic regurgitation.

Classification

I. Level of obstruction

- Valvular – 99%
- Subvalvular and pre-valvular - <1%

II. Severity

Degree	Mild	Moderate	Severe
Aortic valve area sm2	>1.2	0.75-1.2	<0.75
Blood ejection velocity m/s	1-3	3-4	>4

Laboratory and instrumental methods in aortic stenosis diagnosis

ECG:

- left ventricle hypertrophy/overload

X-ray:

- usually insignificant changes of left ventricle size; cardiomegalia is observed in case of heart failure
- calcinosis of aortic valve may be revealed
- poststenotic aorta root dilatation
- in case of congestive heart failure – signs of respiratory congestion

Phonocardiography: confirms auscultation results

Echocardiography with colored Doppler scanning:

- degree of systolic opening of cusps
- location and structure of the leaflets
- aortic valve area
- transvalvular pressure gradient
- thickness of left ventricular wall
- left ventricle function
- differentiation from obstructive hypertrophic cardiomyopathy

Invasive methods are not necessary.

Diagnosis formula:

Includes aethiology and complications.

1. Bicuspidal aortic valve calcinosis. Aortic stenosis. Pulmonary oedema 05.05.98
2. Rheumatic heart disease, inactive phase. Aortic stenosis.

Differential diagnosis:

Other causes of systolic murmur at heart basis:

- increase of blood flow velocity (thyrotixicosis, anaemia)
- obstructive hyperthrophic cardiomyopathy
- pulmonary artery stenosis
- atherosclerotic changes of aortic valve in aged without developing of stenosis

Course and prognosis:

The course is rather favorable. Moderate aortic stenosis doesn't influence on survival; death more often in seventh decade.

Less favorable prognosis after symptoms appearance; the average duration of symptoms (analysis of postmortem examination data; in patients without surgery):

Angina pectoris or syncope – 3 years

Dyspnea – 2 years

Congestive heart failure – 1.5-2 years (cause of death in ½-2/3 patinets).

Sudden death presumably due to arrhythmia – 10-20%

Treatment:

Surgical.

Indications:

- appearance of first symptoms
- severe stenosis even without clinical symptoms

Methods:

- Aortic valve replacement – the most often and effective

Aortic valve replacement is also effective when significant aortic stenosis is complicated by severe left ventricular enlargement. Although the risks of surgery are greater, so are the benefits, and the remarkable improvement in both symptoms and prognosis may follow surgery.

Associated coronary artery disease is usually treated with bypass grafting at the same operation. This combined approach implies that all patients should be studied with coronary arteriography preoperatively, and increases the length of the operation itself.

- comissuritomy

- aortic balloon valvuloplasty, though satisfactory in infants and children, is almost uniformly ineffective in adults in whom the cusps are calcified, and the procedure has been largely abandoned for this age group.

Aortic regurgitation (Aortic insufficiency)

Definition: Aortic regurgitation is a valve disease caused by aortic valve affection or valve ring dilatation causing incomplete cusps closure in diastole and thus blood regurgitation from aorta to left ventricle.

Prevalence

Isolated aortic insufficiency is rather rare valve diseases. It occurs in all age groups with men: women ratio 3:1.

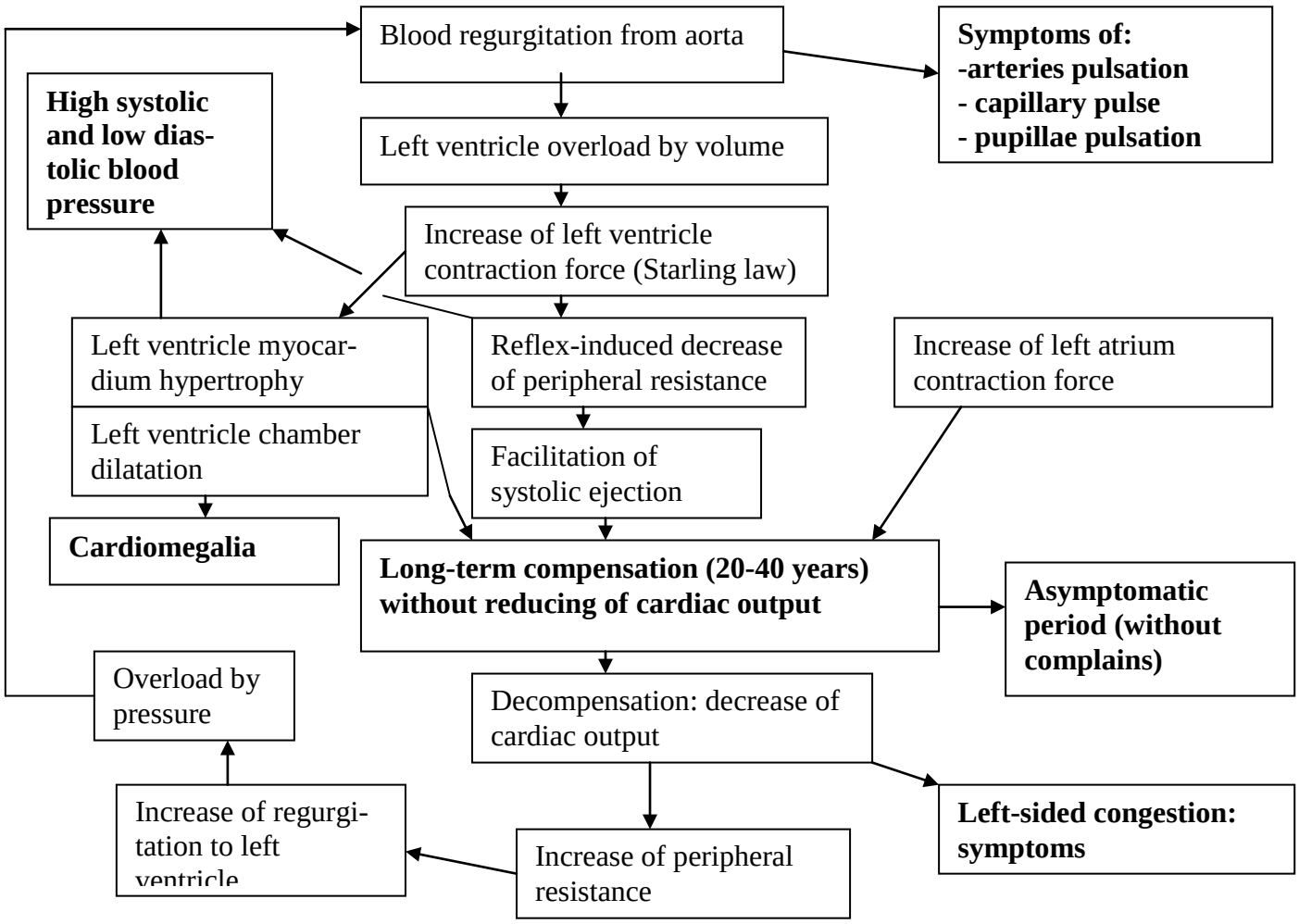
Aethiology:

<table>
<tr><td colspan="2">Chronic aortic regurgitation: 80%</td><td rowspan="2">Acute aortic regurgitation: 20%</td></tr>
<tr><td>Valvular</td><td>Dilatation of aortic valve ring</td></tr>
<tr><td>- rheumatic heart disease
- infectious endocarditis
- congenitally bicuspid valves</td><td>- hypertension
- aortic aneurism
- ankylosing spondylitis
- Marfan’s syndrome
- Reiter's syndrome
- syphilis.</td><td>- rupture (complete or incomplete) of the valve cusp (trauma, infectious endocarditis)
- ascendant aorta dissection</td></tr>
</table>

Morphology:

In case of valve affection: shortening and deformation of cusps with possible calcium deposits (maximal degree of calcinosis – in bicuspidal valve). In case of aortic valve ring dilatation: cusps are usually normal, signs of the main disease are revealed.

Pathogenesis and clinical symptoms:



Acute aortic valve insufficience:

- acute regurgitation: rapid increase of blood volume in left ventricle due to regurgitation from aorta and blood flow from left atrium
- rapid development of pulmonary congestion

Clinical signs of aortic regurgitation depend on aethiology, severity degree, temp of development and compensation mechanisms.

Rheumatic aethiology: the most favorable course: absence of symptoms for 10-20 years; symptoms development at age of 40-50 years old.

Dilatation of aortic ring: more early development of symptoms

Acute regurgitation: first hours/days after defect of leaflet development

Symptoms:

1. **Fatigue**
2. **Brain ischemia:** dizziness, weakness paroxysms
3. **Symptoms due to regurgitation:**

- sensation of severe pushes in chest

- pulsation in head

4. Angina

5. Objective signs

A. due to cardiomegalia

- severe displacement of relative cardiac dullness to the left and downwards
- severe displacement of the apex beat to the left and downwards; apex beat is hyperdynamic and may be sustained

B. due to regurgitation and large stroke volume: vascular symptoms

- jarring of the whole body
- bobbing motion of the head with each systole
- rapidly rising and falling **"water-hammer" pulse**, which collapses suddenly as arterial pressure falls rapidly during late systole and diastole (Corrigan's pulse). This is due to the large stroke volume and rapid diastolic runoff back into the left ventricle
- capillary symptoms – **Quincke's pulse** - alternate flushing and paling of the nail root while pressure is applied to the tip of the nail
- booming "pistole-shot" sound heard over femoral arteries, **double Traube tone**; and to-and fro murmur (**Duroziez's sign**) audible after light compression of femoral artery with the stetoscope
- wide pulse pressure (like 140/60)

C. valve affection symptoms

- SI usually normal
- SII may be markedly reduced (leaflets destruction: infective endocarditis) or increased (luetic aortitis – thickened, dense but movable leaflets)
- SIII, SIV – in case of severe heart failure
- **diastolic murmur:** the earliest and the most typical symptom. Begins with AII (early); decrescendo, pansystolic or lasting up to middle of diastole. Maximally heard in III-IV left intercostals space near sternum; radiating to base of heart and to the apex. If aorta root dilatation is present, the murmur may be heard in II-III intercostals spaces near right border of sternum. It is high-pitched, blowing. Murmur is heard best with the diaphragm of stethoscope, with patient sitting or standing up, leaning forward, with the breath held in forced expiration.
- **Palpable equivalent of murmur: diastolic thrill** along the left sternal border
- **Systolic murmur** at the base of heart due to quick ejection of large blood volume with marked increase of blood flow across the aortic orifice (**midsystolic ejection murmur**)

best heared at the base of the heart, transmitted to the jugular notch and carotid vessels; grade V/VI; higher pitched, shorter and less rasping than that in aortic stenosis.

- **Systolic thrill** in the jugular notch transmitting upward along the carotid arteries
- **Aortic ejection tone**
- **Austin Flint murmur:** soft, low-pitched, rumbling middiastolic or presystolic bruit, probably produced by displacement of the antrerior leaflet of the mitral valve by the aortic regurgitant stream. Earlier onset and longer murmurs correlate with more severe aortic regurgitation.

Acute regurgitation:

- elevation of end-diastolic pressure leading to early closure of the mitral valve (middiastolic sound)
- soft or absent S1
- soft short diastolic murmur
- pulse pressure is not particularly wide

D. Chest deformation: in patients with regurgitation since childhood or youth age: ("cardiac humpback")

6. Left-sided congestive heart failure: usual exertional dyspnea; paroxysmal nocturnal dyspnea and pulmonary edema may also occur.

Classification:

1. **Aethiological (acute and chronic, due to valves affection or valve ring dilatation)**
2. **Degrees:**

	I	**II**	**III**	**IV**
Regurgitation degree (revealed by Doppler)	<20%	20-35%	35-50%	>50%

Laboratory and instrumental signs:

3. ECG: hyperthrophy/overload of left ventricle
4. X-ray:

- severe cardiomegalia due to left ventricle enlargement
- left atrium enlargement may be present
- aorta root, increasing of ascendant aorta may be; dynamic X-ray: increase of ascendant aorta pulsation

5. Phonocardiography: confirms auscultation results
6. Echocardiography:

- changes of valve morphology
- aortic regurgitation degree
- medial leaflet of mitral valve vibration due to mitral and aortic flow
- decrease of mitral valve movement amplitude towards opening
- prematural mitral valve closure
- increase of left ventricle posterior wall and septum movement

Picture description:

Aortic regurgitation is diagnosed by Doppler - most efficiently by Color Doppler Imaging. The regurgitant jet occurs during diastole toward the left ventricular cavity. From the 4 chamber or apical views, the color signal is primarily displayed as red-yellow, since its direction is toward the transducer and the jet can be distinguished from the mitral diastolic inflow.

The long axis view can show the difference between mild regurgitation, which has a narrow base and short jet, compared to severe with broad base and long jet extent. The force in the latter may show presystolic

Diagnostic formula:

Includes main disease, presence of valve disease, complications

1. Rheumatic heart disease, inactive phase. Aortic valve insufficiency.
2. Infective endocarditis. Aortic valve insufficiency. Leaflet rupture. Pulmonary oedema 07.05.97.

Differential diagnosis:

1. Pulmonary valve regurgitation
2. Open ductus arteriosus
3. Sinus Valsava aneurism rupture
4. Arteriovenous fistule

Course and prognosis:

Course is rather favorable. However, when develops, congestive heart failure progresses very rapidly with unfavorable prognosis.

Acute aortic insufficiency is revealed as marked acute respiratory congestion and needs acute surgical treatment.

Treatment:

Surgical (valve replacement).

Acute insufficiency needs emergent surgical treatment.

Chronic course – indications to surgical treatment:

- clinical symptoms appearance

- marked left ventricle dilatation: end0diastolic diameter ≥80 mm and/or end-systolic size≥55 mm.
- some authors also consider ejection fraction <45-50% as the indication for surgical treatment

After-operation prognosis: favorable.

Surgeons are attempting valve repair more frequently in patients with leaflet prolapse (most frequently in individuals with bicuspid valves). Aortic regurgitation due to aortic root disease requires repair or replacement of the root, a more difficult operation.

Right chambers valves affection:

Isolated affection of a.pulmonalis valve and tricuspidal valve are rare (as a possible cause infective endocarditis in narcotic-dependent patients due to unsterile injections conditions). More often relative insufficiency of the valves due to increase of pressure in a.pulmonalis.

Rheumatic Fever and Chronic Rheumatic Heart Disease

Definition

G.Stollerman	V.A.Almazov, E.V.Shlyachto
Rheumatic fever is an inflammatory disease which occurs as a delayed sequel to pharynx-geal infection with group A Streptococci It involves principally the heart, joints, central nervous system, skin and subcutaneous tissues. The usual manifestations in the acute form are migratory polyarthritis, fever and carditis. Sydenham's chorea, subcutaneous nodules and erythema marginatum may oc-cur as other typical manifestations. No single symptom, sign or laboratory test is pathogno-monic of rheumatic fever, although several combinations of them are diagnostic. Although the name acute rheumatic fever emphasizes involvement of joints, rheumatic fever owes its importance to the involvement of heart which can be fatal during the acute stage of the disease and lead to rheumatic heart disease, a chronic condition due to scarring and deformity or the heart valves.	Rheumatic fever is a systemic inflammatory disease of the connective tissue with toxic-immune genesis and with preferable localization of changes in cardiovascular system, which develop as a delayed sequel of an acute β-haemolytocal Streptococci infection group A in pre-disposed to this disease group of people.
Common points: - inflammatory disease - role of group A Streptococcus - heart affection	
Peculiarities	
- Major diagnostic criteria are included into the definition - Peculiarities of clinical course as well as possible outcomes of acute rheumatic fever are mentioned	- Rheumatic fever as a systemic connective tissue disease of toxic-immune origin - One preferable localization (he-art), differentiating it from other diseases of that kind is named - Role of predisposition is underlined

Definition, given by J. M. NEUTZE, emphasizes: the age group, affected by the rheumatic fever, major symptoms and peculiarities of clinical course:

Rheumatic fever is an illness of children and young adults, with major symptoms of arthritis and carditis, a prolonged course, and a tendency to recur. It is the result of abnormal immune reaction to an infection with a Group A β-haemolytic Streptococcus.

Epidemiology

In industrialized countries: 0.07 cases per 1000 per year

In underdeveloped countries: 10 cases per 1000 per year; it remains a major problem; rate of new cases in children and youth is 2.3-3.3%.

The peak incidence is between 5 and 15 years old, rheumatic fever is rare before age 4 and after age 40.

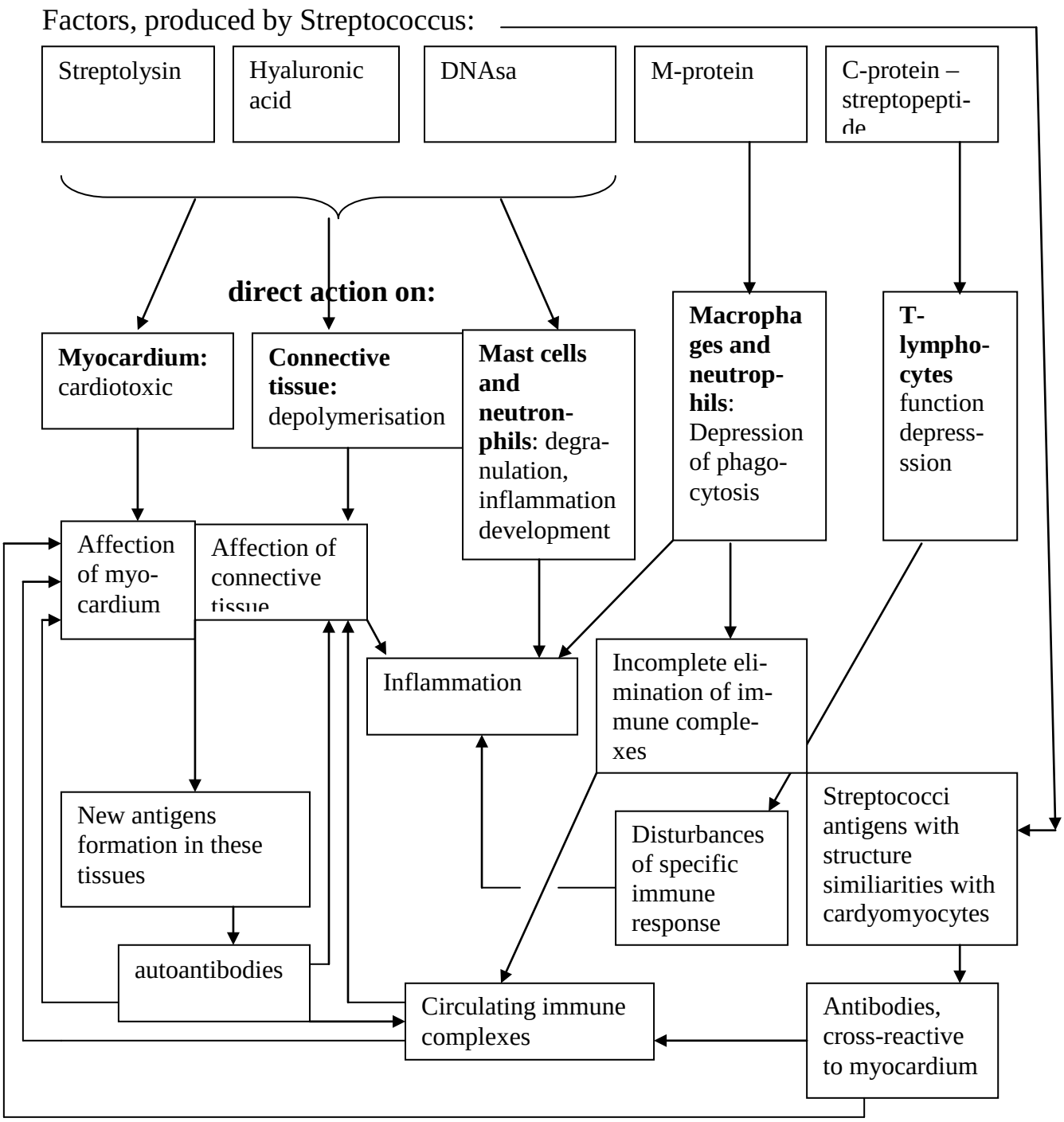
Aetiology and pathogenesis

As it is underlined in all definitions, rheumatic fever is associated with acute inflammation caused by group A β-haemolytical Streptococcus. Usually the inflammation is localized in nasopharyngeal region (pharyngitis, tonsillitis). However, that kind of infection occurs frequently, and rheumatic fever – in 3% and less of the population.

Thus, following factors play a role in disease development:

1. Streptococcus properties: A3, 5, 18, 19, 24 Streptococci; peculiarities of these Streptococci are presence of M-protein and high level of hyaluronic acid.
2. Social and environmental factors: overcrowding (circulation of pathogenic Streptococci strains in population becomes easier), poor housing, poor hygiene; climate (cold, damp weather)
3. Hereditary factor seems to be important, but concrete mechanisms remain unknown. The role of a monoclonal antibody to B cell surface antigens, called D8/17, found in over 90 per cent of patients with rheumatic fever but in only 10 per cent of healthy controls is been discussed.

Pathogenesis



Morphology

The classic histological feature of rheumatic fever is the Aschoff nodule, a perivascular lesion with a central core of necrotic material surrounded by large cells with polymorphous nuclei and basophilic cytoplasm, and an outer layer of lymphocytes.

Picture

Myocardial Aschoff body – the cells are large, elongated, with large nuclei; some are multinucleate

Nodules have a widespread distribution in connective tissues, including those of joints, tendons, and blood vessels. In the heart they are found in myocardial tissue, most valvular lesions consisting of less organized collections of chronic inflammatory cells. The nodules heal by fibrosis, sometimes leading to extensive interstitial myocardial fibrosis. Oedema, collagen fibers frabmentation, fibrinoid necrosis and connective tissue depolimerization are also revealed in myocardium and connective tissue. In joints exccudation usually dominates, significant scarring and deformity are not revealed. Synovitis is usually mild and non-specific.

In 30% of patients first attack is accompanied by valves affection.

The typical valves affection is verrucous valvulitis leading to fibrous thickening, fusion and adhesion of valve comissures and chordae tendineae and thus to various degrees of valvular stenosis and regurgitation. Valve cusps become rigid and deformed.

Aortic valve showing active valvulitis. The valve is slightly thickened and displays small vegetations – "verrucae"

In most of cases mitral valve is affected (75-80%), the structure of the valve becoming severely distorted by progressive fibrosis and eventually calcification; aortic valve is affected less frequently (30%, but rarely as the sole valve), also tricuspid valve can be affected (under 5%); a.pulmonalis valve affection is very rare.

Rheumatic pericarditis produces a serofibrinous effusion with deposit of fibrin on the heart surface. In spite of calcification possibility, pericardial constriction doesn’t occur.

Classification

1. International classification of the diseases:

390 Rheumatic fever without mention of heart involvement

Arthritis, rheumatic, acute or subacute
Rheumatic fever (active) (acute)
Rheumatism, articular, acute or subacute

391 Rheumatic fever with heart involvement

Excludes: chronic heart diseases of rheumatic origin (393-398) unless rheumatic fever is also present or there is evidence of recrudescence or activity of the rheumatic process.

391.0 Acute rheumatic pericarditis

Rheumatic pericarditis (acute)
Any condition in 390 with pericarditis
Excludes: when not specified as rheumatic (420.-)

391.1 Acute rheumatic endocarditis

Rheumatic:
endocarditis, acute
valvulitis acute
Any condition in 390 with endocarditis or valvulitis

391.2 Acute rheumatic myocarditis

Any condition in 390 with myocarditis

391.8 Other acute rheumatic heart disease

Rheumatic pancarditis, acute
Any condition in 390 with other or multiple types of heart involvement

391.9 Acute rheumatic heart disease, unspecified

Rheumatic:
carditis, acute
heart disease, active or acute
Any condition in 390 with unspecified type of heart involvement

392 Rheumatic chorea

Includes: Sydenham's chorea

392.0 With heart involvement

Rheumatic chorea with heart involvement of any type classifiable under 391.-

392.9 Without mention of heart involvement

CHRONIC RHEUMATIC HEART DISEASE (393-398)

393 Chronic rheumatic pericarditis

Adherent pericardium, rheumatic
Chronic rheumatic:
mediastinopericarditis
myopericarditis
Excludes: when not specified as rheumatic (423.9)

394 Diseases of mitral valve

394.0 Mitral stenosis

Mitral (valve) obstruction (rheumatic)

394.1 Rheumatic mitral insufficiency

Rheumatic mitral: Rheumatic mitral:
incompetence regurgitation
Excludes: when not specified as rheumatic (424.0)

394.2 Mitral stenosis with insufficiency

Mitral stenosis with incompetence or regurgitation

394.9 Other and unspecified

Mitral (valve): Mitral (valve):
disease (chronic) failure

395 Diseases of aortic valve

Excludes: when not specified as rheumatic (424.1)

395.0 Rheumatic aortic stenosis
Rheumatic aortic (valve) obstruction
395.1 Rheumatic aortic insufficiency
Rheumatic aortic:
incompetence
regurgitation
395.2 Rheumatic aortic stenosis with insufficiency
Rheumatic aortic stenosis with incompetence or regurgitation
395.9 Other and unspecified
Rheumatic aortic (valve) disease
396 Diseases of mitral and aortic valves
Involvement of both mitral and aortic valves whether specified as rheumatic or not
397 Diseases of other endocardial structures
397.0 Diseases of tricuspid valve
Tricuspid (valve) (rheumatic):
disease
obstruction
stenosis
397.1 Rheumatic diseases of pulmonary valve
Excludes: when not specified as rheumatic (424.3)
397.9 Rheumatic diseases of endocardium, valve unspecified
Rheumatic:
endocarditis (chronic)
valvulitis (chronic)
Excludes: when not specified as rheumatic (424.9)
398 Other rheumatic heart disease
398.0 Rheumatic myocarditis
Excludes: myocarditis not specified as rheumatic (429.0)
398.9 Other and unspecified
Rheumatic:
carditis
heart disease NOS
Excludes: carditis not specified as rheumatic (429.8)
heart disease NOS not specified as rheumatic (429.9)

2. Working group classification (used in Russia)

<table>
<tr><td>Phase</td><td colspan="2">Clinical anatomical characteristics</td><td>Course (for active)</td></tr>
<tr><td>Active (I, II, III activity degree) – corresponds to active rheumatic fever or recurrent rheumatic fever</td><td>Rheumocarditis:
- primary without valves affection
- recurrent without valves affection
- without visible signs of heart affection</td><td>- arthritis
- serosites
- chorea
- ehcephalitis
- vasculitis
- skin affection
- nephritis</td><td rowspan="2">- acute (all signs of activity of disease disappear in up to 3 months)
- subacute (3-6 months)
- prolonged (more than 6 months)
- recurrent (constant exacerbations)
- latent (minimal clinical and laboratory signs of activity)</td></tr>
<tr><td>Inactive (corresponds to chronic rheumatic heart disease)</td><td>- cardiosclerosis
- valves affections</td><td>-</td></tr>
</table>

Activity degrees
III degree:
Active clinical manifestations
- ESR >40 mm/hr
- marked increase of C-reactive protein, fibrinogen, α2- globulin, antistreptococcus antibodies
II degree: less marked changes
I degree: minimal signs

Clinical features

1. Infection preceding the disease
Some two-thirds of patients give a history of prior sore throat, usually 1 to 3 weeks before the development of rheumatic symptoms.

2. Intoxication syndrome

Fever, varying from patient to patient, weakness, perspiration, appetite disorders. However, subacute course may occur with less marked intoxication. In some cases no acute bout is recognized at all, the patient presenting with established rheumatic heart disease.

3. Arthritis - in 30-60% of patients

- affection of most commonly affects larger joints, especially wrists, elbows, knees, and ankles.
- Classical signs of arthritis include redness, warmness and swallowing of joints; however sometimes objective signs are usually limited to minor warmth and swelling, but pain may be excruciating, especially with pressure or movement.
- Migration of arthritis: one joint will be affected for 2 to 3 days and then the inflammatory process moves to another region, two or more joints may be affected simultaneously to some degree.
- Arthralgia without objective signs may occur in other joints or may be the only feature, symptoms varying from minor discomfort to severe pain; myalgia may be also present.
- Quick positive effect of non-steroid anti-inflammatory drugs is typical
- Untreated, joint pains usually settle over 1 to 4 weeks.

4. Carditis in almost all cases

A. MYOCARDITIS:

in adults and youth its course is usually mild; severe course is more often in children

- **pain:** usually non-intensive, that can't be concretely defined and described by patient (cardialgia)
- signs of **contractile function disturbances** varying from subclinical, which are most often (very mild dyspnea, tachycardia, hypotonia) to the marked signs of congestive heart failure syndrome (in children)

- **objective signs of heart muscle affection:** cardiomegalia, decreased intensity of the heart sounds (decrease grade depends on severity), systolic murmur over the heart apex (in mild course – not-intensive, due to papillary muscles dysfunction, in severe course with marked heart enlargement – more intensive); III and IV sounds.
- **Arrhythmic syndrome** may also occur, usually as supraventricular and ventricular ectopic beats.

Picture :

a:Chest radiograph of an 8 year old patient with acute carditis before treatment (heart chambers enlargement, signs of congestion in lungs)
b. Same patient after 4 weeks of treatment (disappearance above mentioned signs)

B. PERICARDITIS

- chest pain: rather intensive, constant, with aggravation during inspiration

- objective signs: superficial scratchy sound of the pericardial bruit. A moderate pericardial effusion may develop, marked effusion usually doesn't occur.

C. VALVULITIS

Is usually diagnosed retrospectively, after several months, when the valve disease develops. The clinical features depend on the valve affected and grade of affection.

5. Chorea

Once present in 50 per cent of patients, Sydenham's chorea is now recognized in about 5 per cent. It has a longer latent period than arthritis or carditis, from 1 to 6 months. It may occur in association with carditis, or as the only rheumatic manifestation.

Chorea is characterized by:

- jerky, purposeless movements, exaggerated by tension but disappearing in sleep. Clumsiness, grimacing, emotional lability, and unclear speech may appear and, in severe cases, violent movements and progressive weakness develop.
- Occult chorea may be diagnosed by testing sustained hand grip. When the patient holds the observer's fingers, minor sudden movements and fluctuations in muscle tension can be detected.

Symptoms usually subside over 1 to 3months but may persist for a longer period and occasionally recur over the following 2 years. They may occasionally reappear during the administration of oral contraceptives or during pregnancy.

6. Erythema marginatum and rheumatic nodules

These manifestations are seen much less frequently but may contribute to diagnosis.

- **Erythema marginatum** begins as a non-itchy, faint red macule, the erythema spreading outward while the centre returns to normal colour. The margin is often irregular in outline, and adjacent areas may coalesce. The rash usually fades in 24 h but may recur over a period of months. *Erythema marginatum also occurs in association with acute glomerulonephritis and drug reactions, and occasionally without recognized cause.*

- **Rheumatic nodules** are more common in children. They are firm, painless, movable, nontender, 0.5 to 2 cm in diameter, and situated over tendons or bony prominences over extensor surfaces of the joints, particularly knees, wrists and elbows. They persist for weeks and are recurrent. Subcutaneous nodules are rarely seen and when present, they are usually associated with severe carditis.

7. Other symptoms. Other organs involvement:

- epistaxis (quite common)
- vasculitis (rare)
- nephritis (rare)
- fibrinous pleurisy (rare); even more rare pleural effusion, as a rule, associated with pericardial effusion
- rheumatic pneumonitis (rare)
- encephalitis (rare)

Changes, revealed by laboratory and instrumental methods:

1. **Inflammatory changes in blood:**
- ESR rise
- Leucocytosis with increase of neutrophils' percentage
- C-reactive protein - positive
- Elevated α_2 - and γ- globulins levels
- Elevated fibrinogen level

2. **Signs of Streptococcus infection:**
- β-haemolytical Streptococcus presence in pharynx, revealed by microbiological methods
- antibodies revealed to Streptococcus factors (Antistreptolysin O – ASO, rise of the level over 250; Antistreptokinase, Antihyaluronidase – AH – rise of the level over 300; AntiDNAse B)

3. **Signs of immune system reaction**
- increase of circulating immune complexes level
- decrease of complement level in blood

4. **Signs of heart affection**

- **Cardiospecific enzymes'** rise may occur in cases of severe myocarditis
- **ECG** The P-R and Q-T intervals may be prolonged on the electrocardiogram, non-specific changes of ST and T can be found (usually biphase or isoelectric or negative T); supraventricular and ventricular ectopic beats may be present. If valves affection is present, ECG reveals overload of respective heart camera.
- **X-ray:** cardiomegalia; in severe cases – changes, typical for congestive heart failure; in case of valves affection – typical configuration changes (mitral etc)

- **Echocardiogram:** left ventricle hypokinesis, efection fraction lowering, mitral regurgitation. In case of valves affection – verrucous valvulitis, valve cusp thickening with decreased opening in stenosis, regurgitation also may be revealed, specific chamber enlargement can be seen.In case of pericardium effusion fluid is revealed in pericardium cavity.

Diagnosis

1. Diagnosis is based on Jones Criteria:

Major criteria	Minor (additional) criteria
- Carditis - Polyarthritis - Chorea - Erythema marginatum - Subcutaneous nodules	- fever - arthralgia - previous rheumatic or rheumatic heart disease - elevated ESR or positive C-reactive protein - prolonged QT interval

1. **Presence of two major or one major and two minor criteria indicate a significant basis for diagnosis statement**.
2. **Carditis** diagnosis is based on following evidences:

- Pericarditis.
- Cardiomegaly, detected by physical signs, radiography, or echocardiography
- Congestive failure, right- or left-sided
- Mitral or aortic regurgitation murmurs, indicative of dilation of a valve ring with or without associated valvulitis.
- The Carey-Coombs short middiastolic mitral murmur may be present.

In the absence of any of the above definitive signs, the diagnosis of carditis depends upon the following less specific changes:

- Electrocardiographic changes: The most significant is PR prolongation greater than 0.04 s above the patient's normal. Changing contour of P waves or inversion of T waves is less useful.
- Changing quality of heart sounds.
- Sinus tachycardia persisting during sleep and markedly increased by slight activity.
- Arrhythmias, shifting pacemaker, or ectopic beats.

Course and prognosis:

In cases if valves are not affected, the course of the disease is rather favorable. If valves changes are present, recurrent attacks are usually present. The most severe exacerbations are observed in patients with prosthetic valves.

Recurrences of rheumatic fever are most common in patients who have had carditis during their initial episode and in children, 20% of whom will have a second episode within 5 years. Recurrences are uncommon after 5 years and infrequent in patients over 25 years of age.

Following types of course may be present: acute, subacute, prolonged, relapsing and latent (in last case no signs of rheumatic fever are revealed in history, only the manifestations of rheumatic heart disease are seen as valves affection. According to some data, rheumatic heart disease is revealed in patients without rheumatic fever history in about 40% of cases) .

Valves affection:

- In 70% the first bout ends without valves affection (reconvalescence)
- In 30% valves are affected; in case of more acute rheumatic fever course valves insufficiencies are formed, in less activity of inflammation – stenoses are more usual.
- After 10 years of disease, two-thirds of patients have detectable valvular affection.

Each recurring attack increases the risk of valves affection or progression of preceding valve affection. Progression of valves affection and recurrent myocarditis bouts lead to progressive heart failure.

Prognosis

Initial episodes of rheumatic fever may last months in children and weeks in adults. The immediate mortality rate is 1–2%. Persistent rheumatic carditis with cardiomegaly, heart failure, and pericarditis imply a poor prognosis; 30% of children thus affected die within 10 years after the initial attack. Eighty percent of affected children attain adult life, and half of these have little if any limitation of activity.

In developing countries, acute rheumatic fever appears earlier in life, and the evolution to chronic valvular disease is accelerated.

Differential diagnosis

1. With non-infectious and non-immune diseases, affecting heart

- thyrotoxicosis
- vascular dystonia
- alcohol disease, alcohol myocardiopathia
- silent myocardium ischemia

In these cases only heart affection is present; no inflammatory changes in blood, no signs of Streptococcus infection and immune response to it are observed in these cases.

2. With autoimmune diseases (heart, joints affection, possible intoxication syndrome, inflammatory signs in blood)

- Lupus erythematosus

- Scleroderma
- Rheumatoid arthritis etc

Course of arthritis, character of skin lesions, more common kidneys involvement, comparatively less often valves affection. No signs of Streptococcus infection are revealed.

3. Infective endocarditis: heart (including valves), joints affection, skin changes, intoxication syndrome, inflammatory changes in blood.

Especially difficult situation is when endocarditis develops in patient with rheumatic heart disease. Presence of Streptococcus infection laboratory signs from one side and revealed bacteriemia help in these cases.

The most difficult the differential diagnosis is in patients with rheumatic heart disease and heart failure; in these cases biochemical inflammatory signs may be absent, the only one manifestation of the disease exacerbation may be progression of heart failure or appearance/progression of rhythmus and conductivity disorders.

Treatment

A. General Measures:

Bed rest should be enforced until: return of temperature to normal without medications; normal sedimentation rate; normal resting pulse rate (< 100/min in adults); and return of ECG to baseline. Patient should be treated in a hospital.

B. Medical Measures:

1. Salicylates:

- markedly reduce fever and relieve joint pain and swelling.
- have no effect on the natural course of the disease.
- the duration of treatment is usually 6-8 weeks

Doses:

Adults:

- Aspirin, 0.6–0.9 g every 4 hours (4-6 g daily);
- Diclophenac (150 mg daily)
- Nimesulid and Meloxicam (selective cyclooxygenase inhibitor)

Children are treated with lower doses.

Side effects: the most usual – non-steroid anti-inflammatory drugs related gastropathy varying from minimal signs to stomach mucosa erosions, acute ulcers and bleeding. In selective cyclooxygenase inhibitors, these effects are significantly less marked.

2. Penicillin (anti-Streptococcus treatment)

Benzylpenicilline – 6 mln units daily (1mln units x 6 times a day i.m.)

Cephalosporines can be used instead.

The course of treatment is 2 weeks.

3. Corticosteroids–

administered in following cases:

- no effect is shown by non-steroid anti-inflammatory drugs
- severe myocarditis course
- active rheumatic attack in patients with prosthetic valves

Dose: initially 40-60 g with gradually reducing after inflammatory process decrease (course is usually described as 2-weeks long), after cessation – 2-3 weeks of non-steroid anti-inflammatory drugs.

Prevention of Recurrent Rheumatic Fever

- The initial episode of rheumatic fever can usually be prevented by early treatment of streptococcal pharyngitis.

Prevention of initial attacks of RF (primary prevention) requires the eradication of GAS from the pharynx. Table 3 shows the currently recommended treatment schedules.[42]

Emphasis should be given to the need of eradication of GAS as part of the treatment of acute RF.

Primary prevention of rheumatic fever.

Agent	**Therapeutic scheme**
Benzathine penicillin G	600,000 U for patients < 27Kg; 1,200,000 U for patients > 27kg, IM (once) Or
Penicillin V	Children: 250mg 2-3 times daily, PO (10 d) Adolescents: 500mg 2-3 times daily, PO (10 d)
For individuals allergic to penicillin:	
Erythromycin: Estolate Ethylsuccinate	20-40mg/kg/d 2-4 times daily, PO (10 d) or 40mg/kg/d 2-4 times daily, PO (10 d) (maximum 1g/d)

The following are unsuitable: sulfonamides, trimetroprim, tetracyclines and chloranphenicol.

Antibiotic prophylaxis is the safest way to prevent recurrent attacks of acute RF and is recommended for patients with well-documented RF. The recommendations from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular

Disease in the Young of the American Heart Association see next table

Secondary prevention of rheumatic fever.[42]

Agent	**Therapeutic Scheme**
Benzathine penicillin G	1,200,000 U every 4 weeks*, IM or
Penicillin V	250mg twice daily, PO or
Sulfadiazine	500mg once daily for patients < 27kg; 1g once daily for patients > 27kg, PO
For individuals allergic to penicillin and sulfadiazine:	
Erythromycin	250mg twice daily, PO

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

* In Russia and some other countries, Bicillin-5 1.2 mln units is also used i.m. every month (5 years, in some countries – during rest of the life) is used.

** Sulfonamides or Erythromycin: If the patient is allergic to penicillin, sulfadiazine (or sulfisoxazole), 1 g daily, or erythromycin, 250 mg orally twice daily, may be substituted.

Chorea treatment:

Helpful in decreasing the severity of involuntary movements but may not improve the behavioral symptoms:

- haloperidol (initial dose of 0.5 to 1mg/kg/day, maximum, 5mg/day) [49]
- valproic acid (15-20 mg/kg/day)

May be also used:

- Carbamazepine
- Alternative - Phenobarbital also may be used, 5-7mg/kg/day, tid. Treatment is usually maintained for 8-12 weeks.