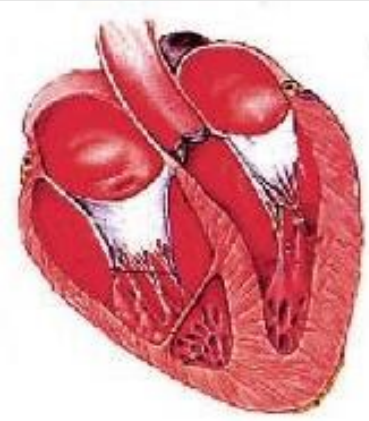
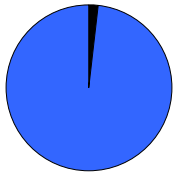
CONGENITAL AND ACQUIRED VALVULAR HEART DISORDERS

Dr. Pavel Maruna

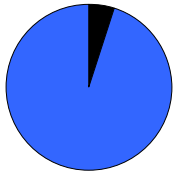


Valvular heart disease

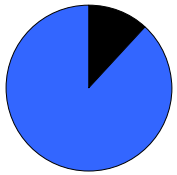
Prevalence of moderate or severe valvular disorder



2 % …in total population



4 - 5% … above 65 yr.

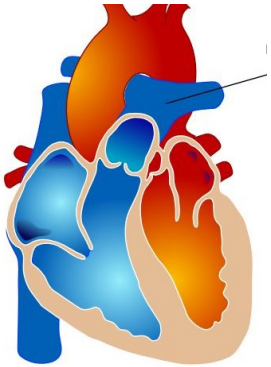


12% … above 75 yr.

Occurrence of valvular disorder → 1,5x risk of death

Valvular heart disease

Health risk



Cardiac failure

Bacterial endocarditis

Atrial fibrillation

Pulmonary embolism

Stroke

15x

How to diagnose valvular disease?

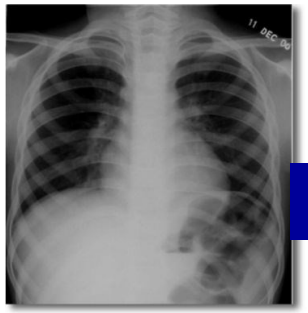
Physical examination

murmur

Usually years before first clinical manifestation

How to diagnose valvular disease?

ECG

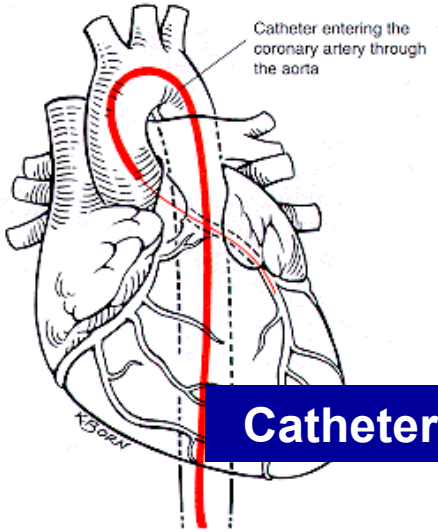


X-Ray

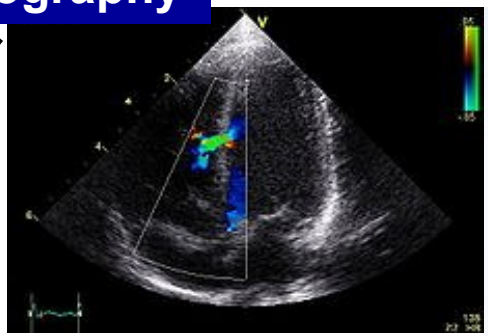
Physical examination

murmur

Usually years before first clinical manifestation

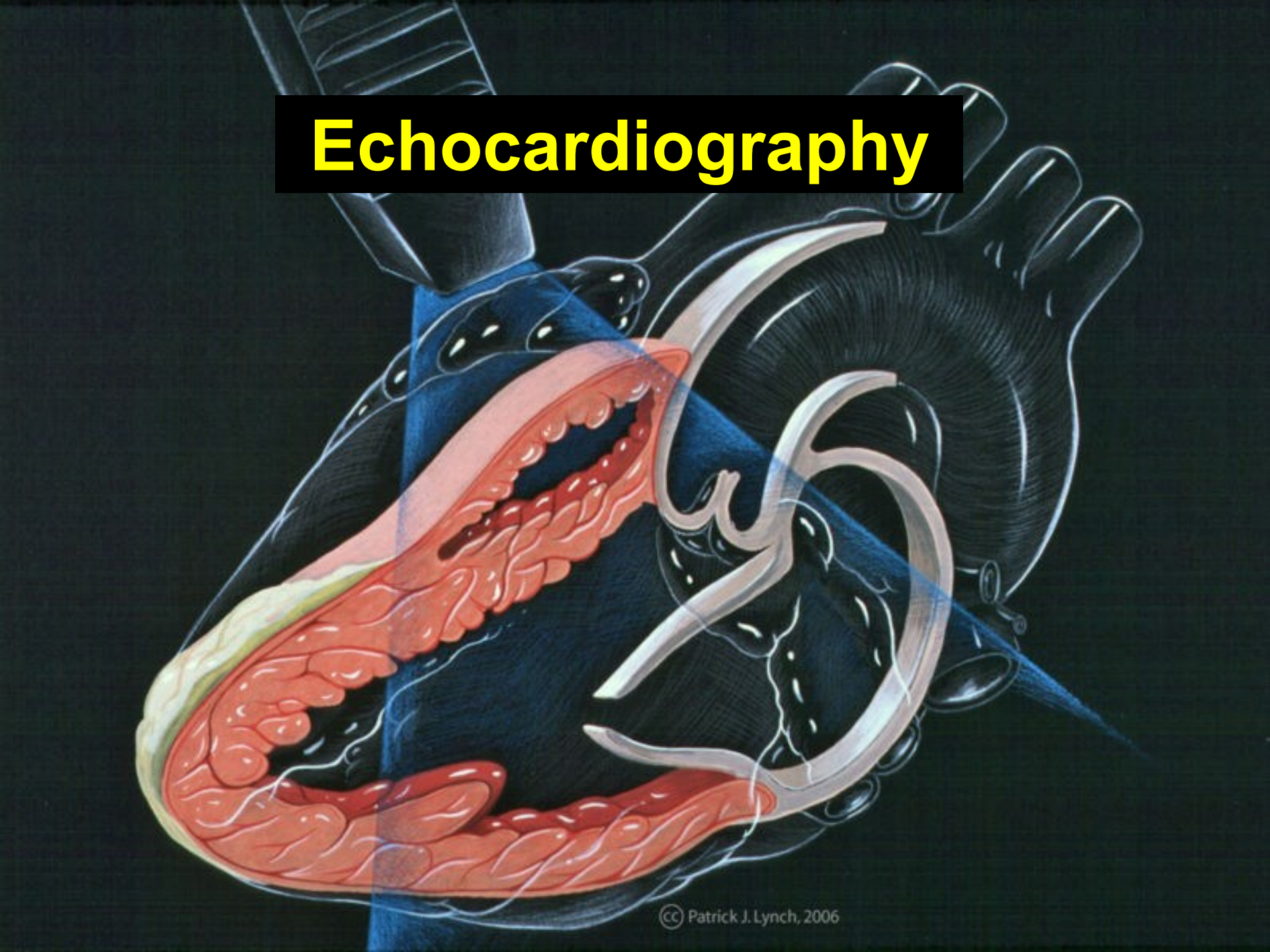


Echocardiography



Catheterization

Echocardiography

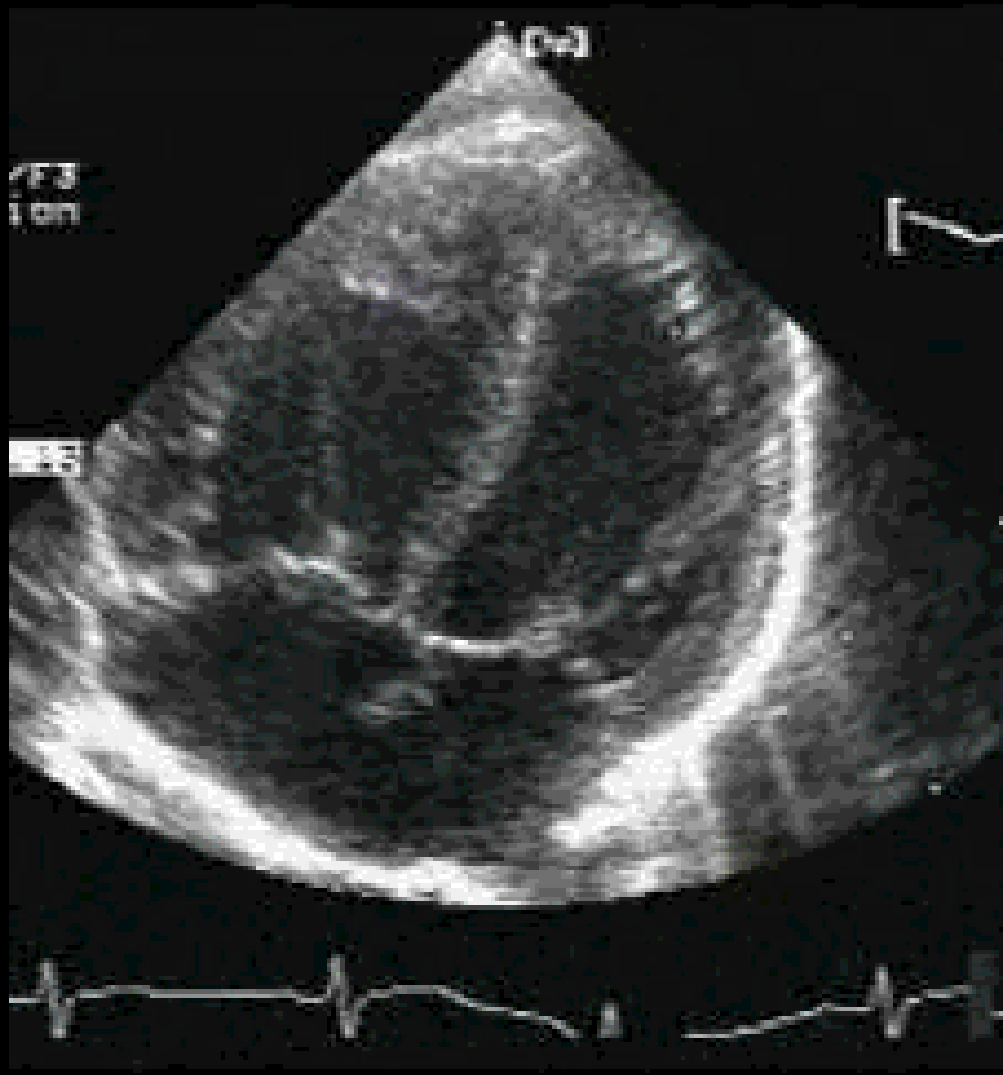


Echocardiography

2D image

1D (Doppler)

2D + color Doppler



Echocardiography

2D image

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Echocardiography

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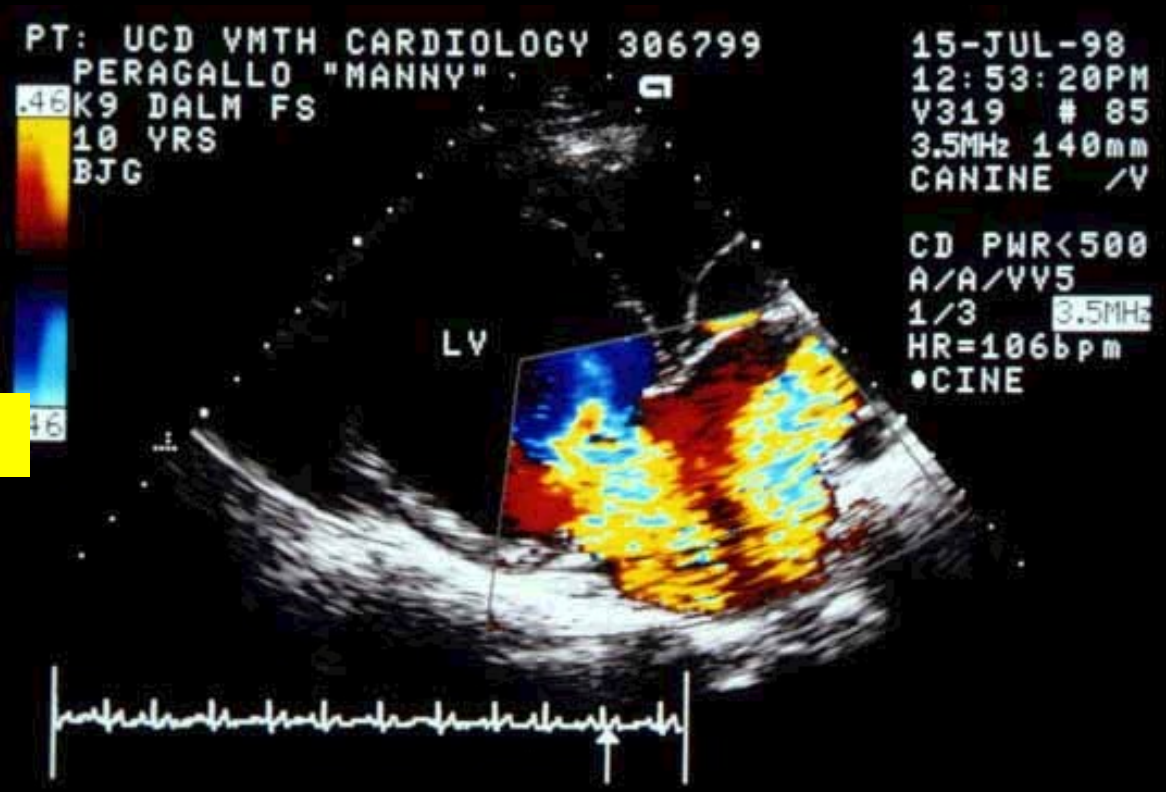


Catheterization

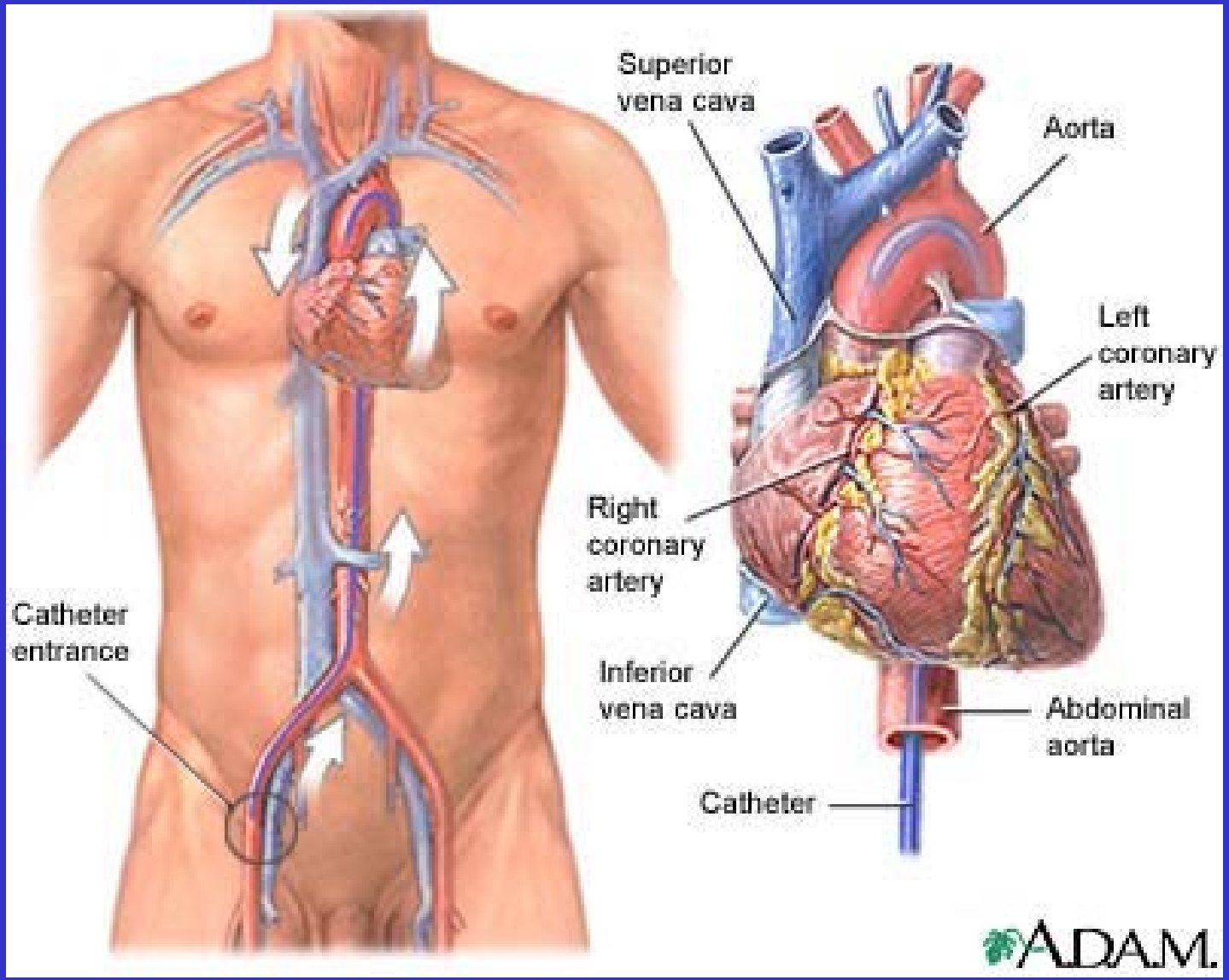
Invasive measurement (not only) of BP

a. femor. – aorta – LV
v. femor. – v. cava inf. – RV

- pressure measurements in separate heart cavities
- wedge pressure – end-diastolic pressure
- pressure gradients
- cardiac output
- blood for oxygen saturation
- injection of contrast dyes for angiography
- biopsy

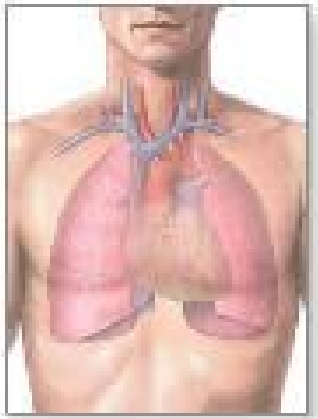
Catheterization

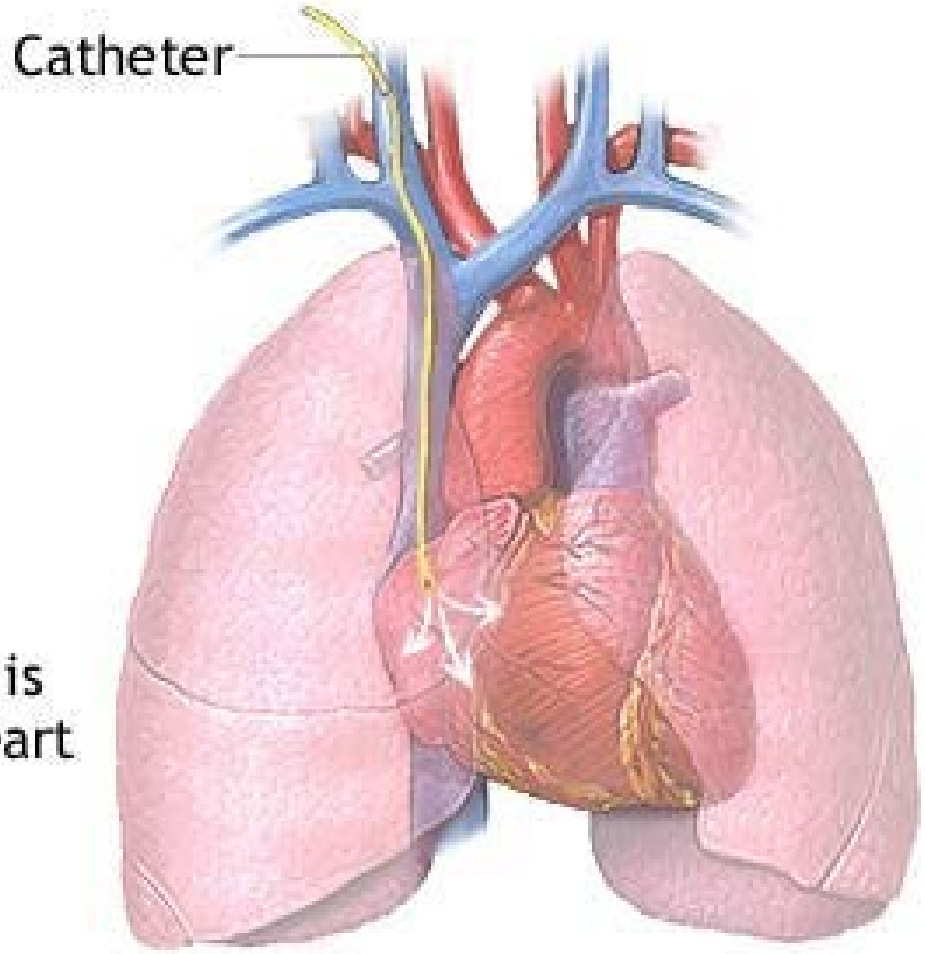
Left heart catheterization (A. femoralis – aorta)



Catheterization

Right heart catheterization (V. femoralis - V. cava – RA – LV – A. pulmonaris)

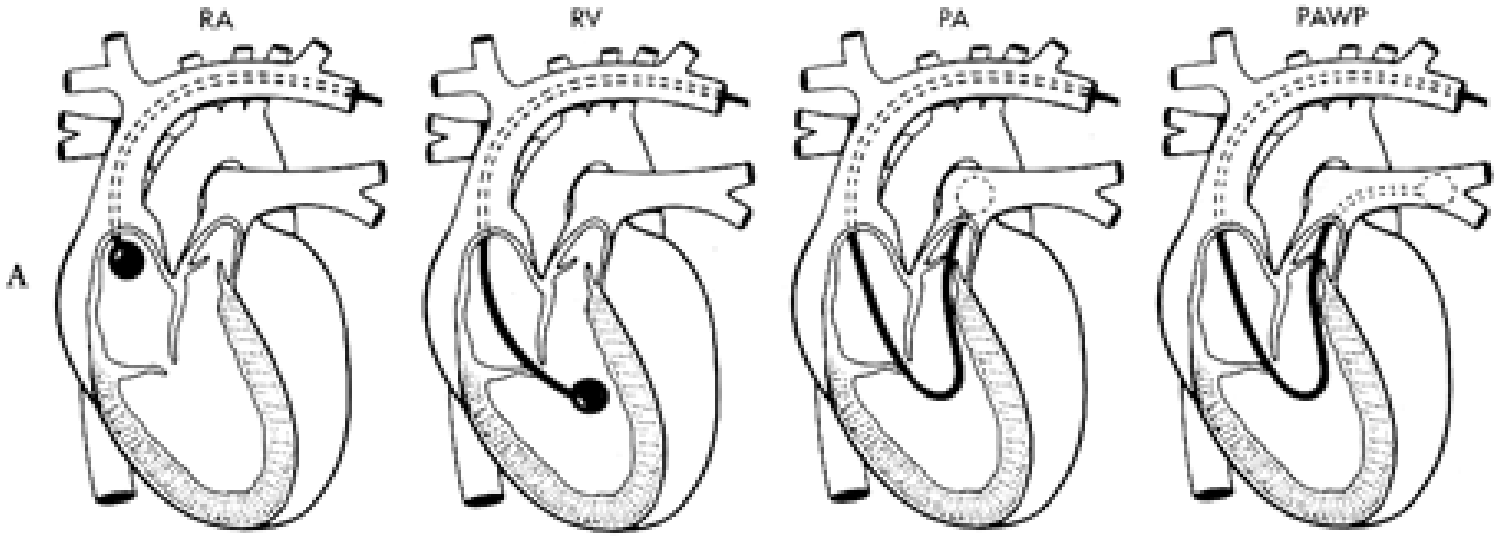




Heart catheterization

Swan-Ganz catheter position in the heart

- Right atrium (RA)
- Right ventricle (RV)
- Pulmonary artery (PA)
- Pulmonary artery wedge pressure (PAWP)



Brief historical overview

16th to 19th century — **First attempts of cardiac surgery were associated with high mortality…**

Human sacrifice with heart extraction in Aztec culture

Brief historical overview

1895

The **first cardiac surgery**
Norwegian surgeon Axel Severin Capellen, Kristiania (now Oslo).

Ligation of bleeding coronary artery in a 24 yr. old man in a deep shock.

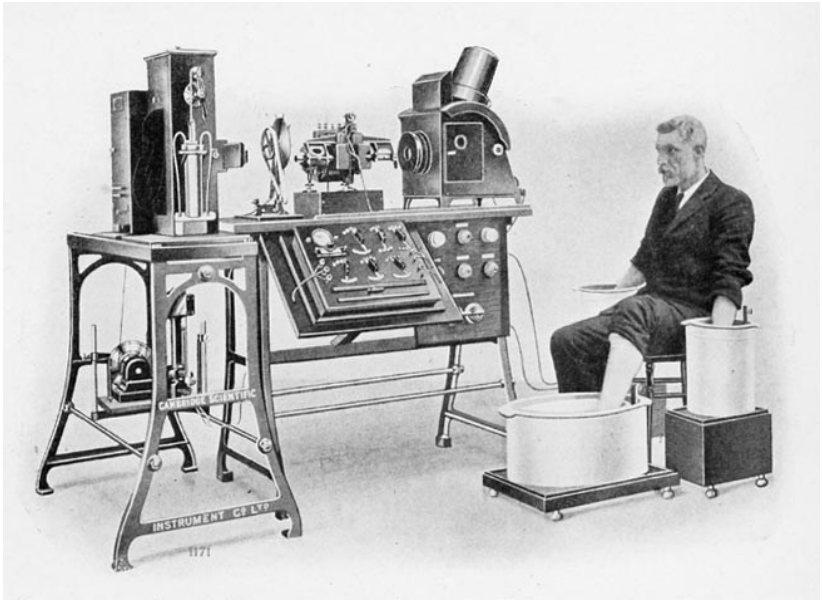
Patient died after 24 hours

Brief historical overview

1895

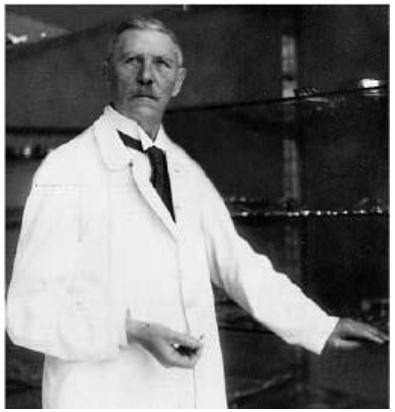
Electrocardiography

Willem Einthoven
The first accurate ECC recording
(including names for P, Q, R, S and T waves)



Brief historical overview

1896



The first successful heart surgery.
Dr. Ludwig Rehn, Frankfurt, Germany,
who repaired a wound to the right ventricle

Brief historical overview

1925 The first **surgery of valvular disease**.

Dr. Henry Souttar operated successfully on a young woman with mitral stenosis.

He cut left atrium and inserted a finger into this chamber … to palpate and explore the damaged mitral valve.

The patient survived for several years

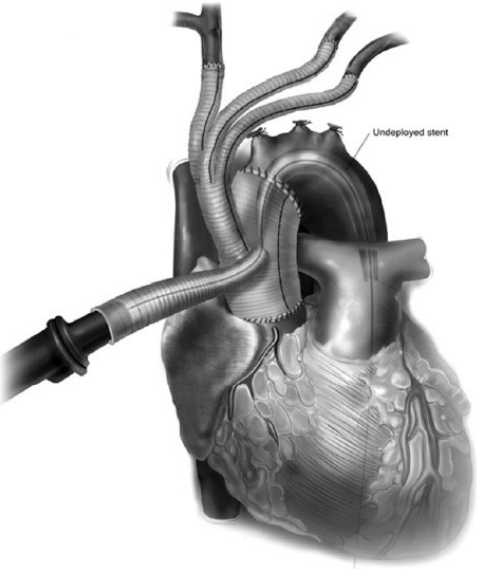
Brief historical overview

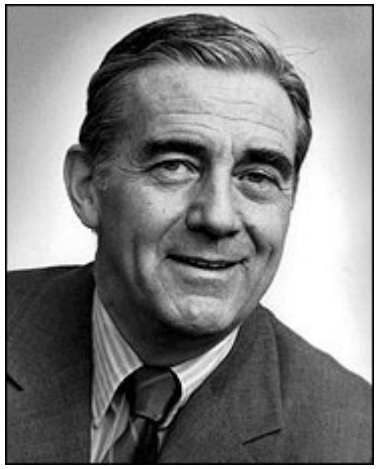
1929 **The first heart catheterization**
German physician Werner Forssmann.

A plastic tube was inserted in cubital vein and guided to the right heart ventricle … under x-ray control.

Since the late 1970s, heart catheterization has been extended to diagnostic and therapeutic uses
… as a less invasive treatment of valvular diseases and ischemic heart diseases

Brief historical overview



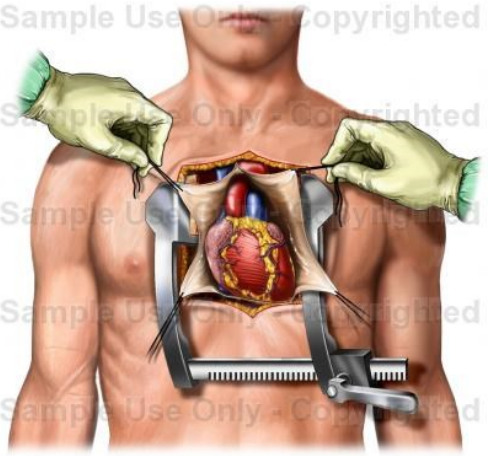


Dr. Wilfred Gordon Bigelow,
University of Toronto, Canada

During the surgery, the heart is exposed and the patient's blood is send to cardiopulmonary bypass.

1952 **The first surgery on open heart**

The method allows to repair intracardial abnormalities including valvular diseases

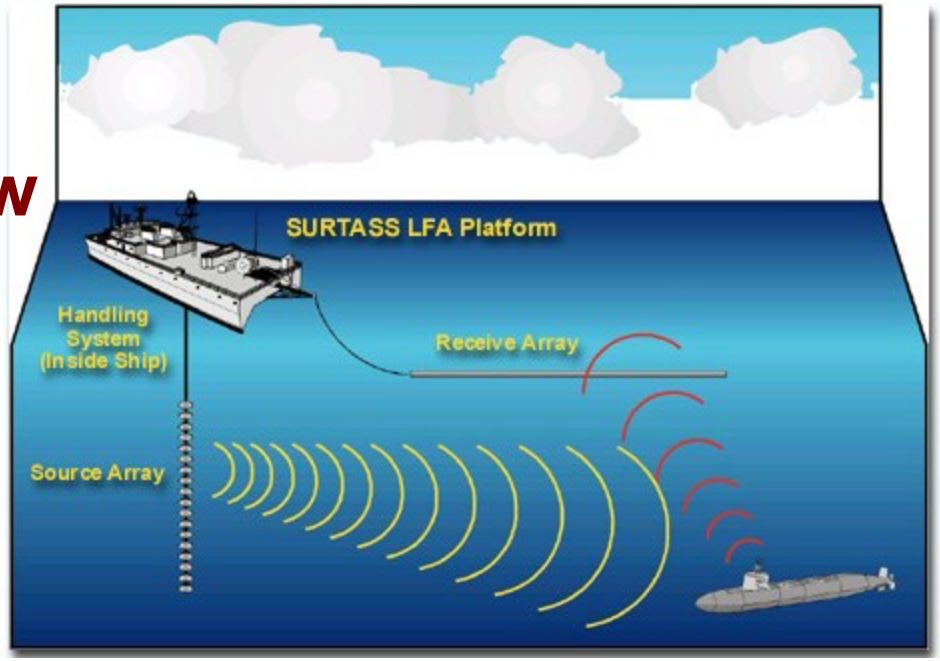


Brief historical overview

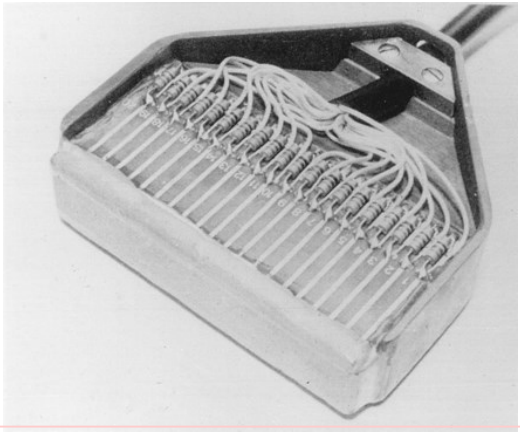
1952 **The first cardiac surgery of congenitgal heart defect in a deep hypothermia**

Dr. C. Walton Lillehei and Dr. F. John Lewis
University of Minnesota, USA

Brief historical overview



Sonar in WW2
– predecessor of diagnostic ultrasonography



„Phonograph“ – ultrasonography in 50s years

1957 Echocardiography in cardiological diagnostics
J. J. Wild and H. D. Crawford, USA

Pathogenesis of valvular disorders

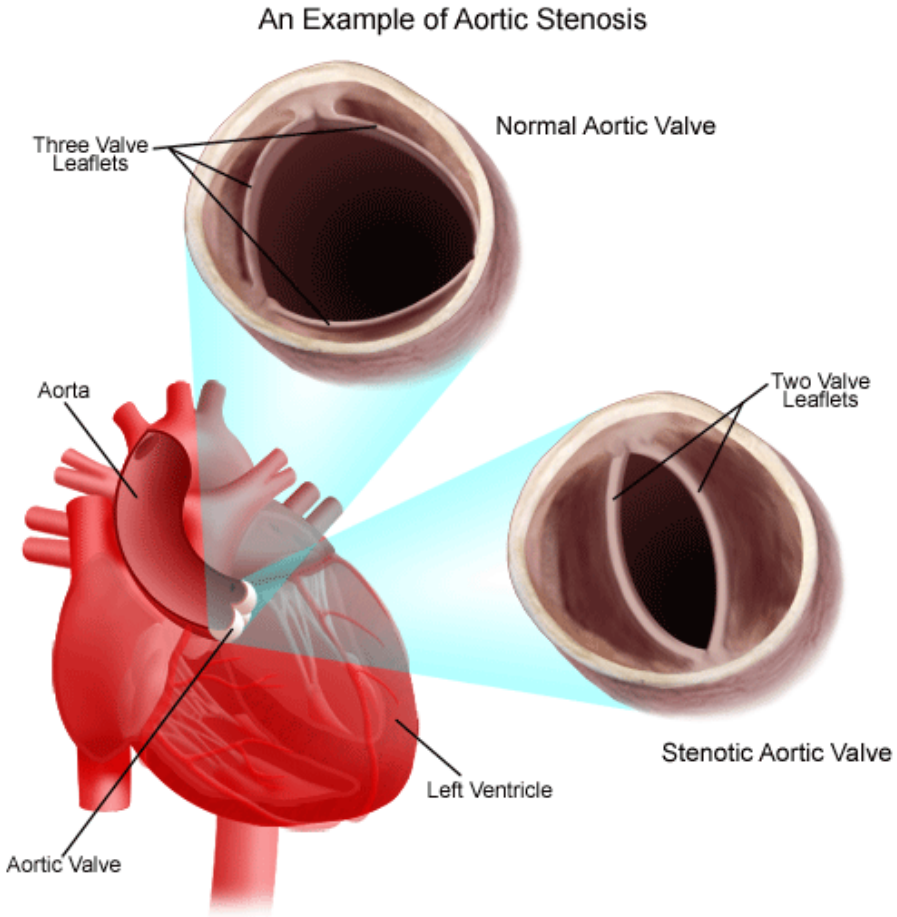
Etiology:

- **rheumatism**
- **infectious endocarditis**
- **ischemic heart disease**
- **pox**
- **valvular degeneration**

…

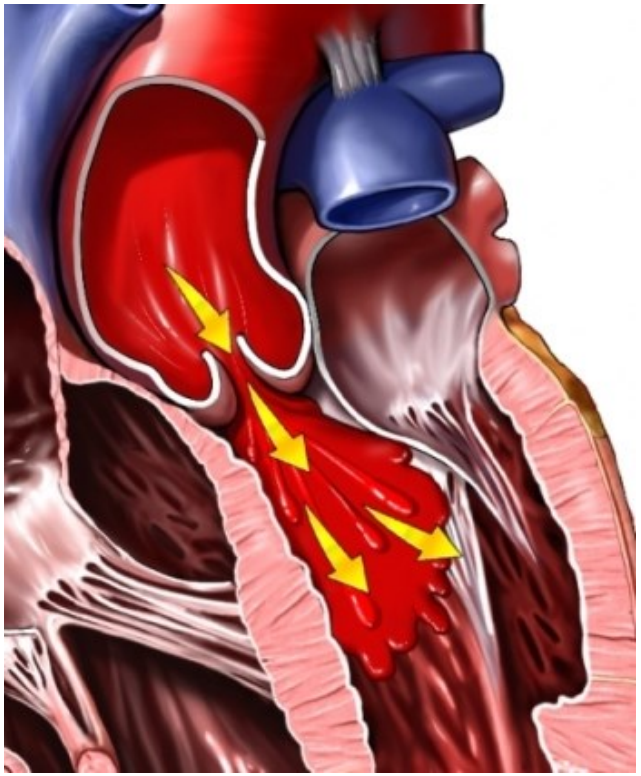
Pathogenesis of valvular disorders

Stenosis = impaired opening of a valve



Pathogenesis of valvular disorders

Insufficiency = impaired closure of a valve, allowing (hemodynamically important) back blood flow



Pathogenesis of valvular disorders

Course

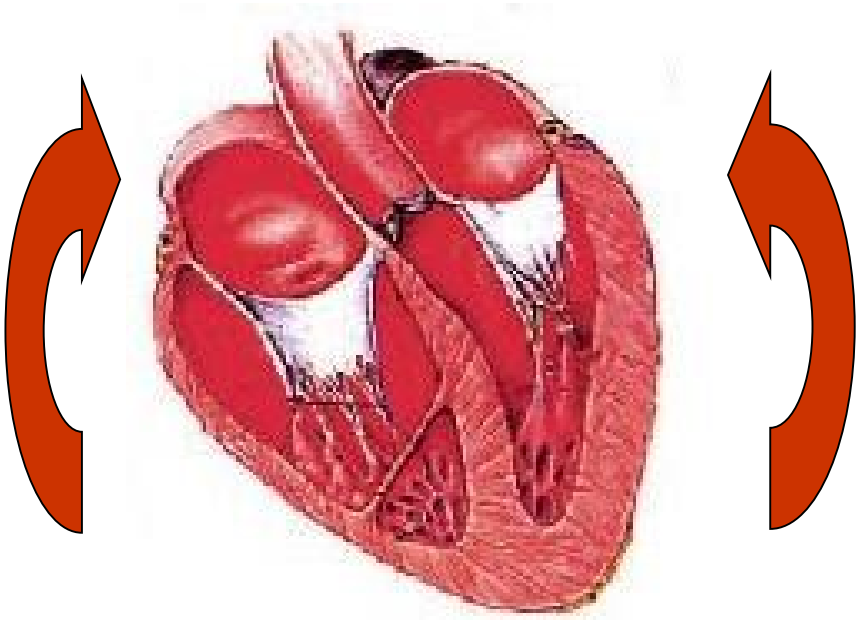
Long latent development (often 10x years)

Terminal heart failure

Hemodynamic impact

„Retrograde“ progression influencing a pulmonary circulation and a right heart

„Tricuspidalisation“



Venous tone
Breathing
Muscle „pump“

Venous return

Preload

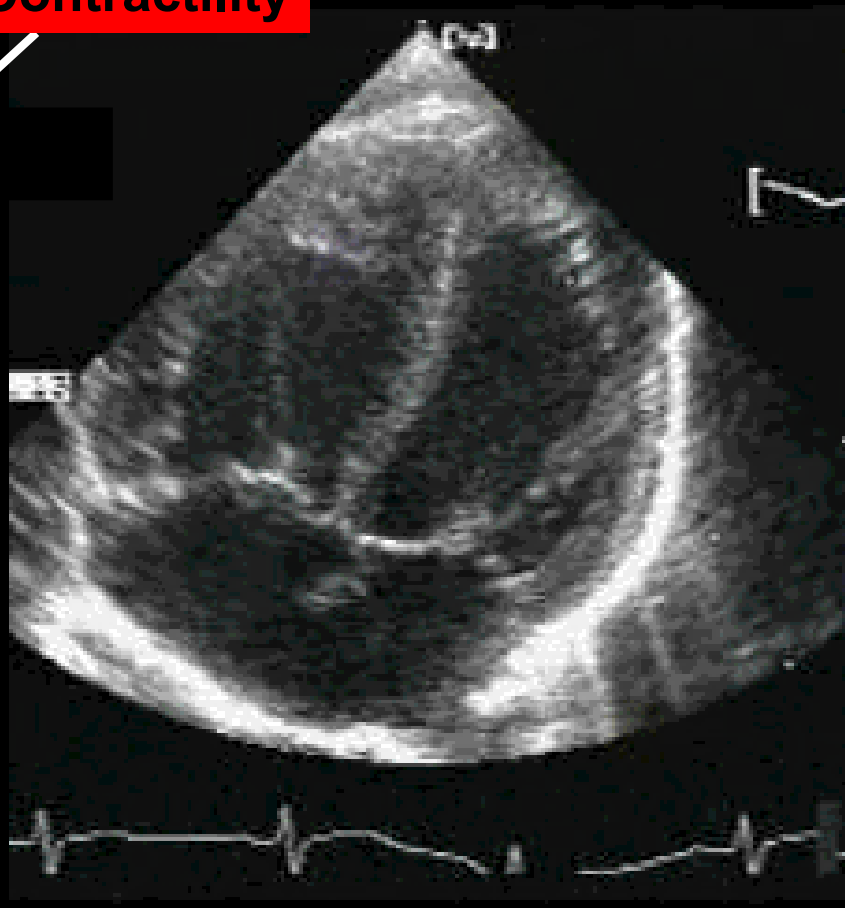
Catecholamines
Ischemia,
inflammation
Cardiotonic drugs

Contractility

Stroke volume

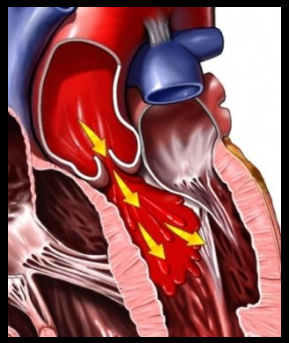
Afterload

Peripheral resistance



Venous tone
Breathing
Muscle „pump“

Venous return



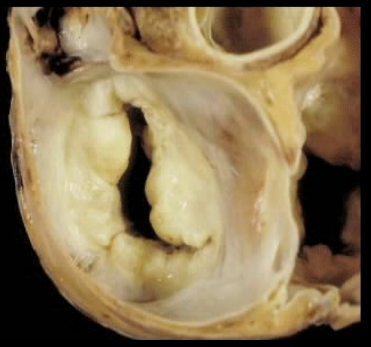
Valvular insufficiency

Preload

Stroke volume

Afterload

Peripheral resistance

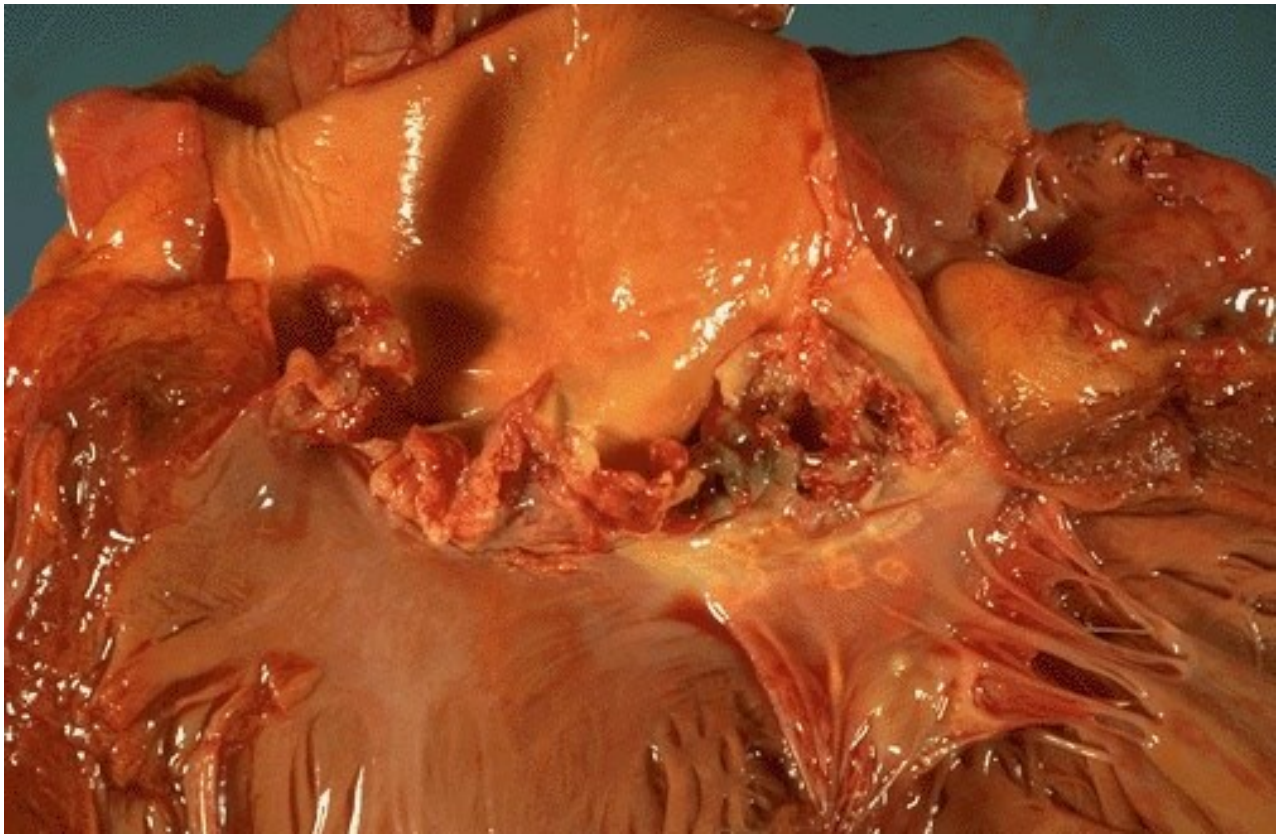


Valvular stenosis

Pathogenesis of valvular disorders

Other complications

Bacterial endocarditis (endocarditis maligna lenta)



Pathogenesis of valvular disorders

Other complications

Bacterial endocarditis

Arrhythmia (atrial fibrillation)

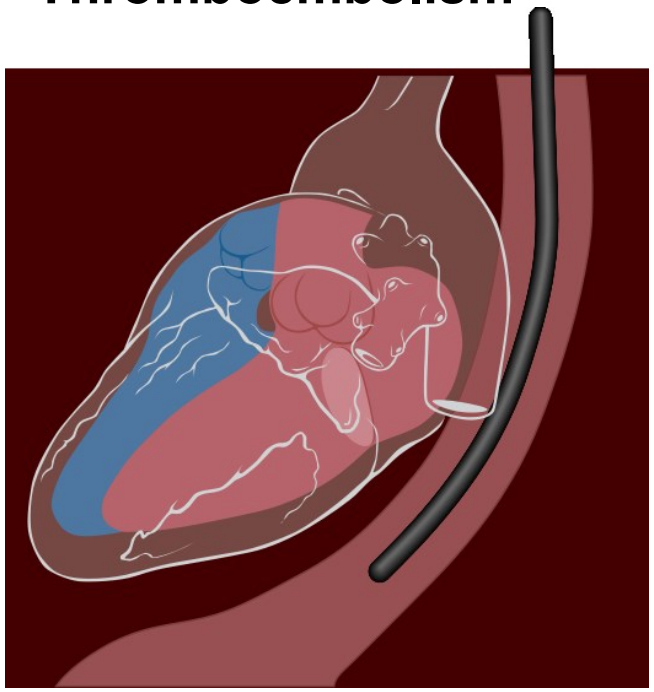
Atrial activity = irregular *f* waves

Completely irregular transmission to ventricle

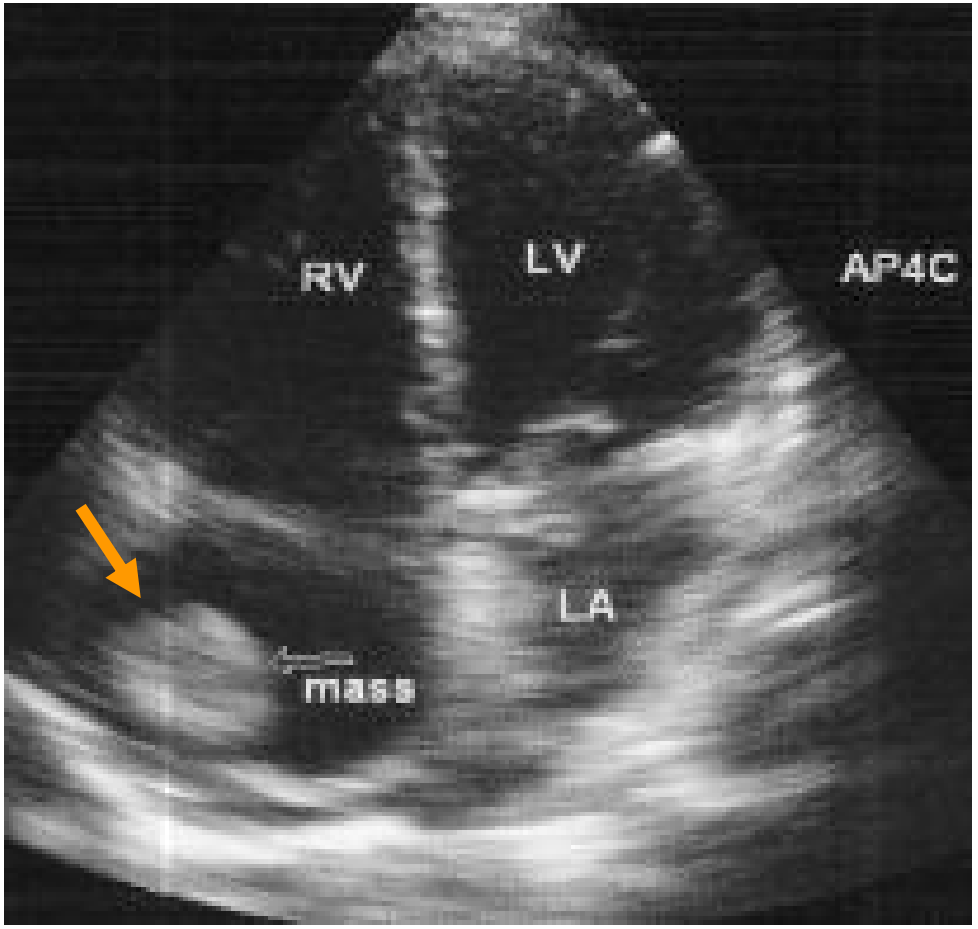
Pathogenesis of valvular disorders

Other complications

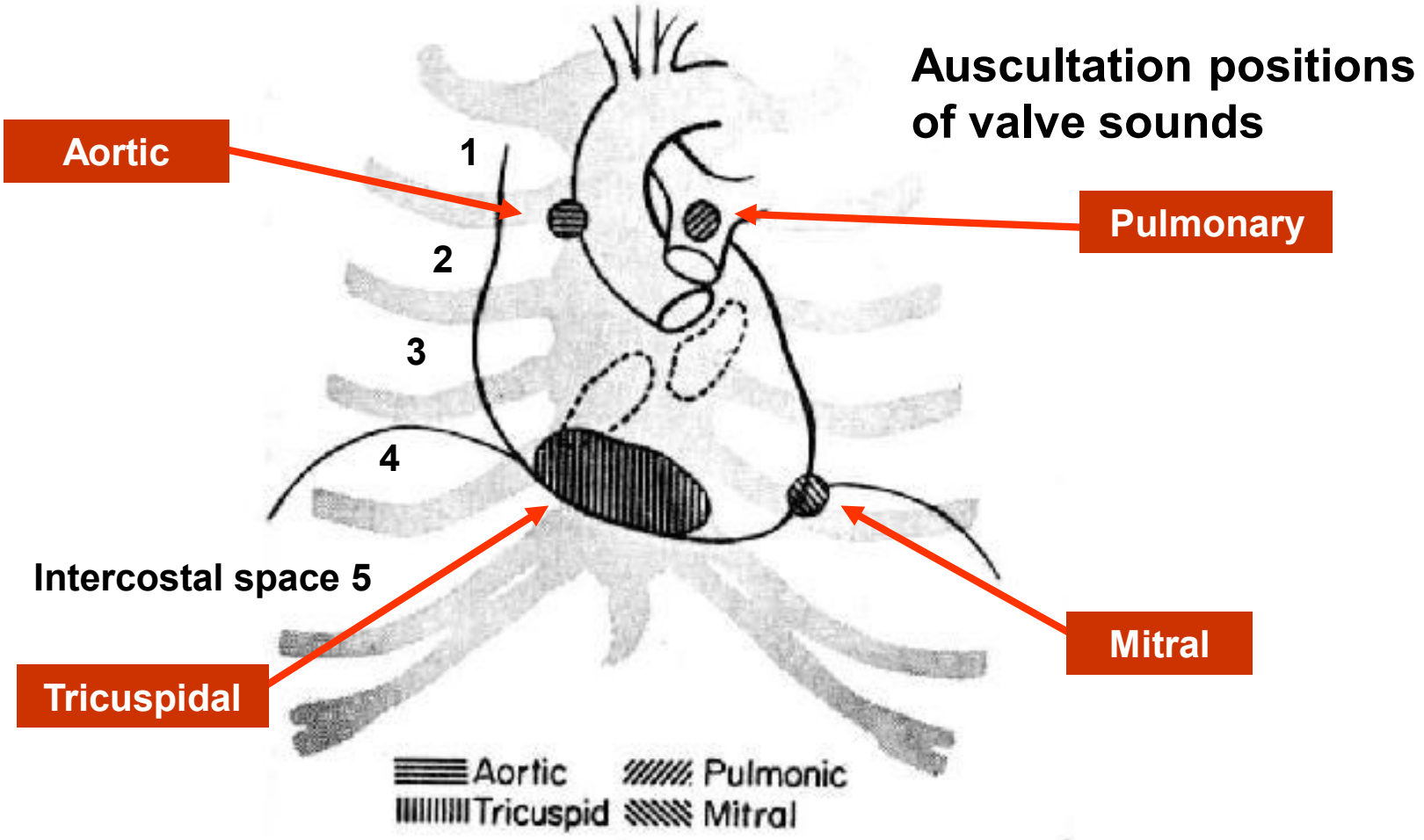
Bacterial endocarditis
Arrhythmia
Thromboembolism

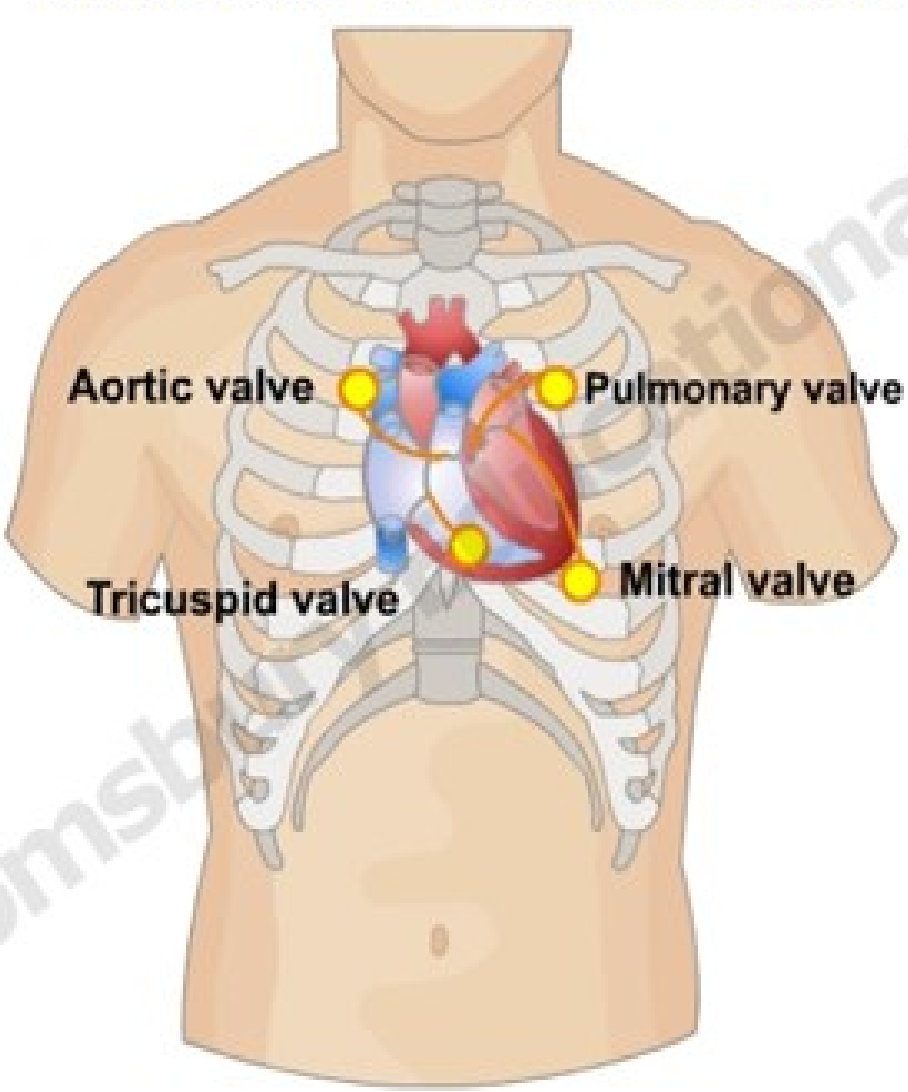


TEE

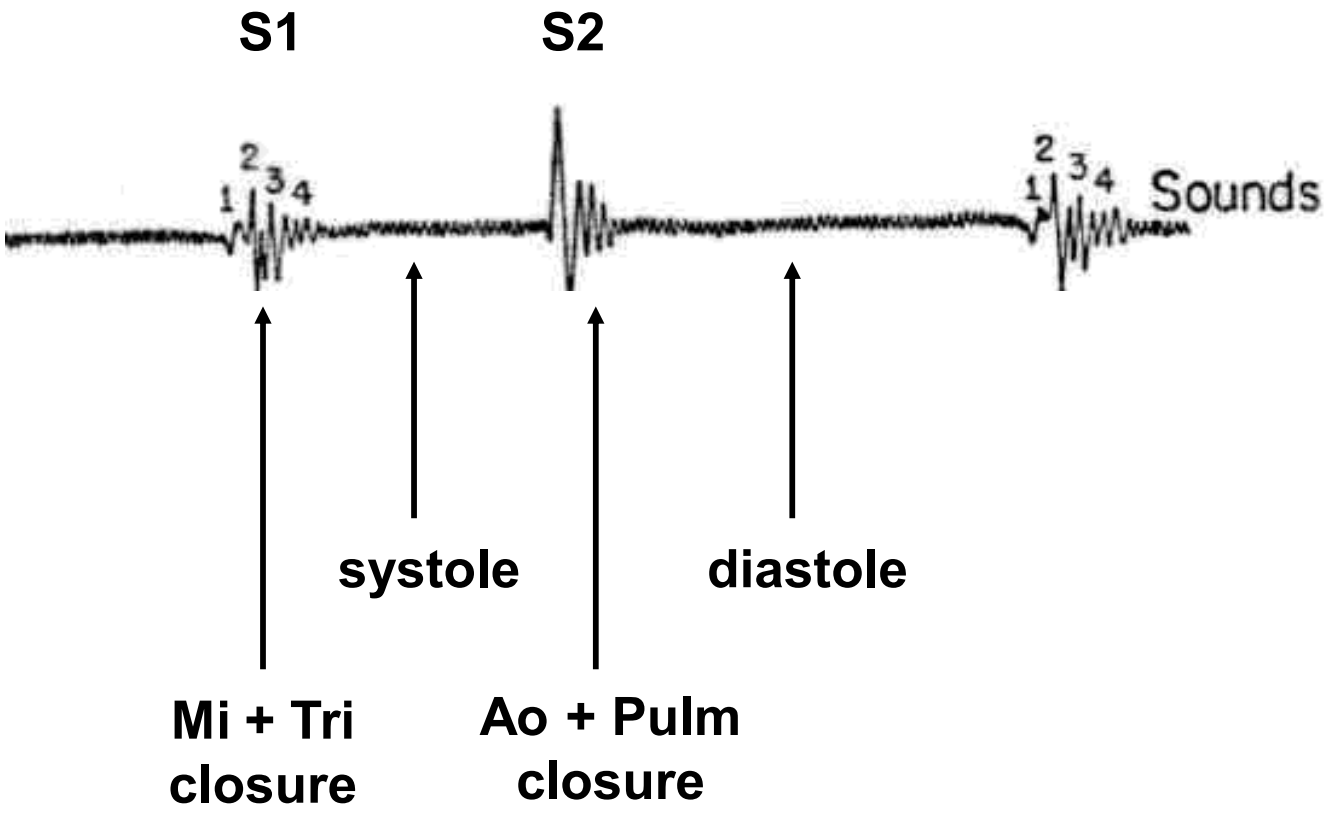


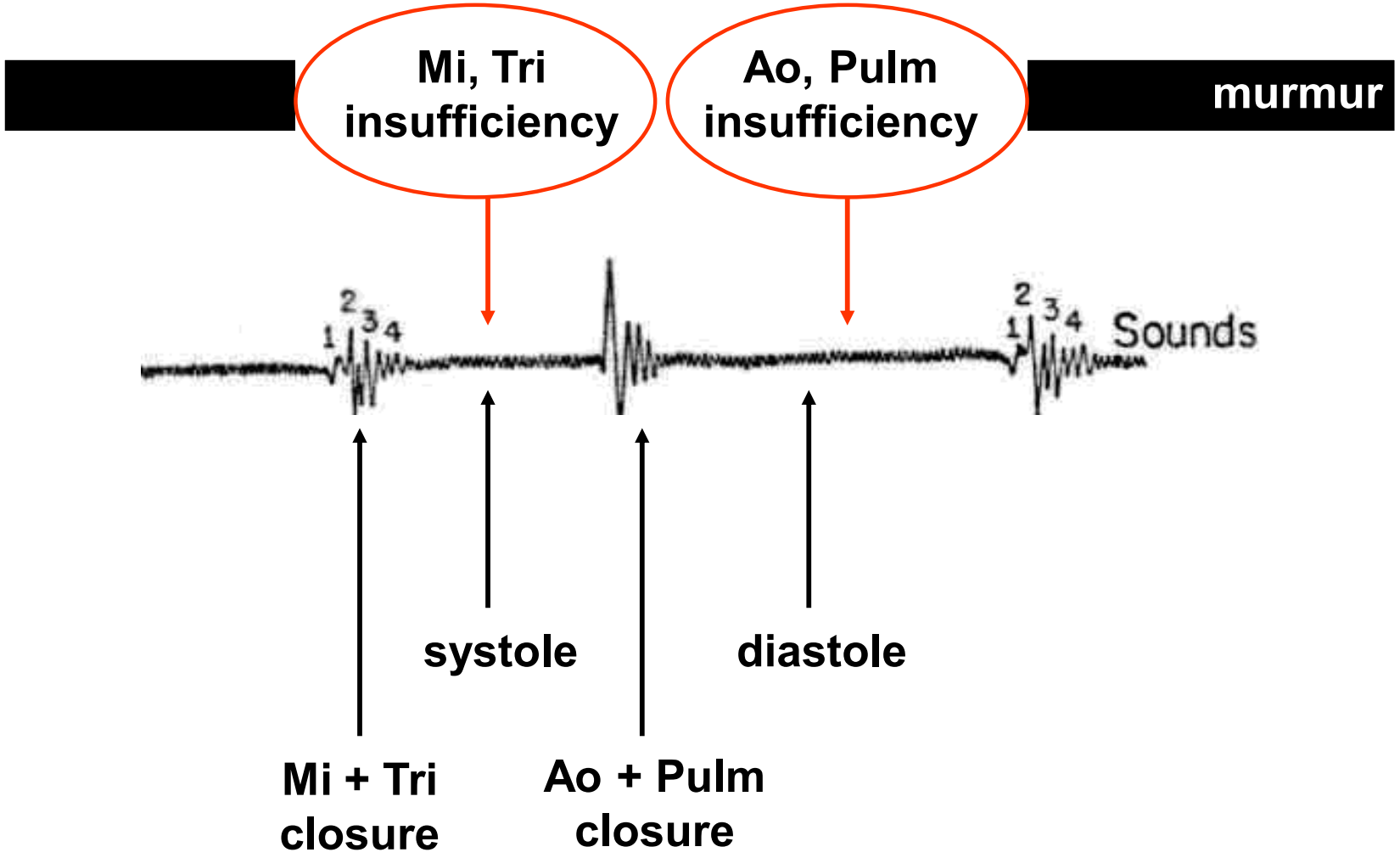
Auscultation

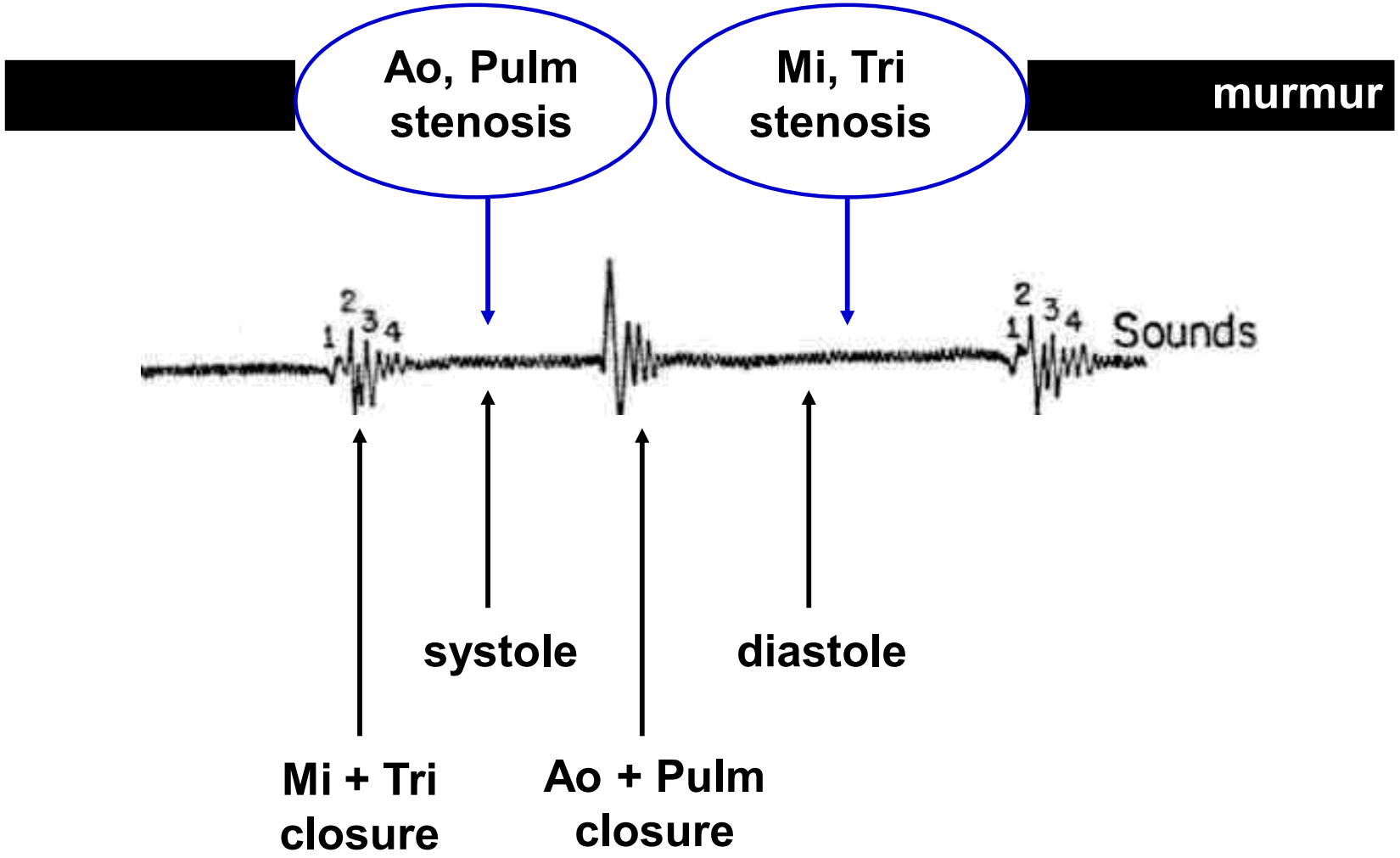



Aortic valve
Pulmonary valve
Tricuspid valve
Mitral valve

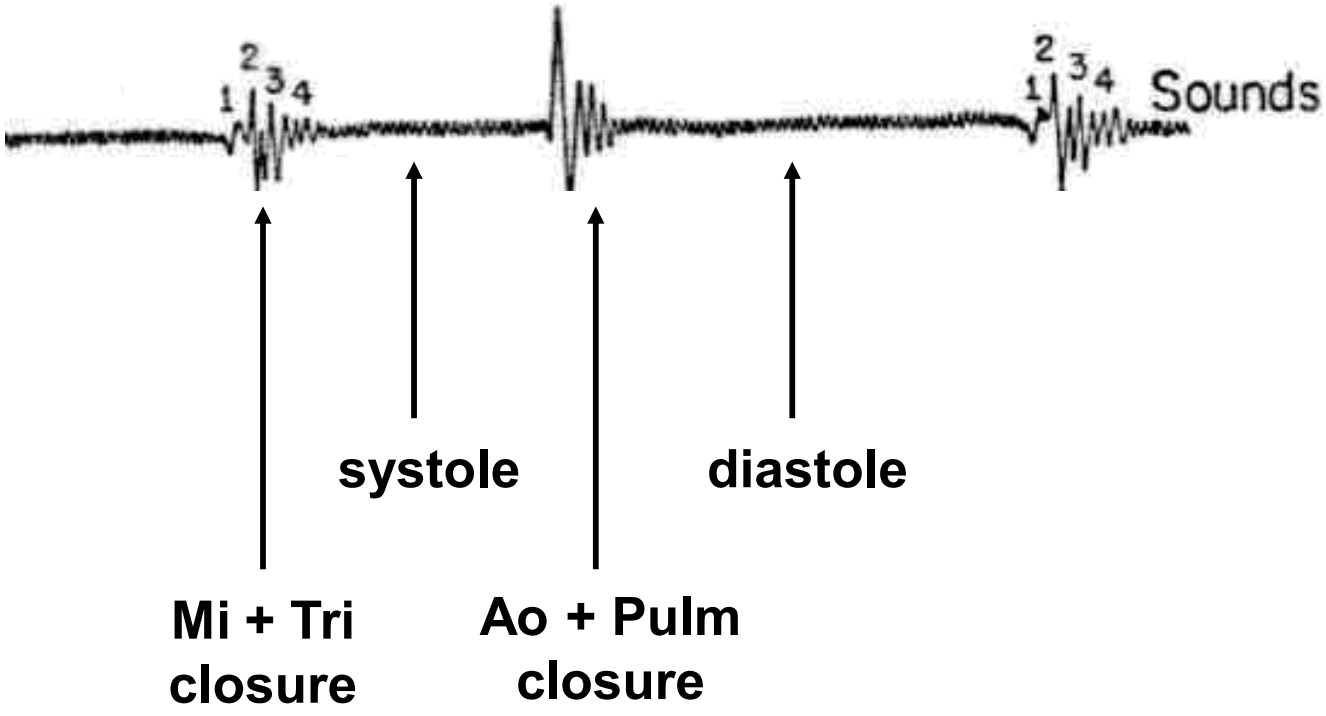
Auscultation



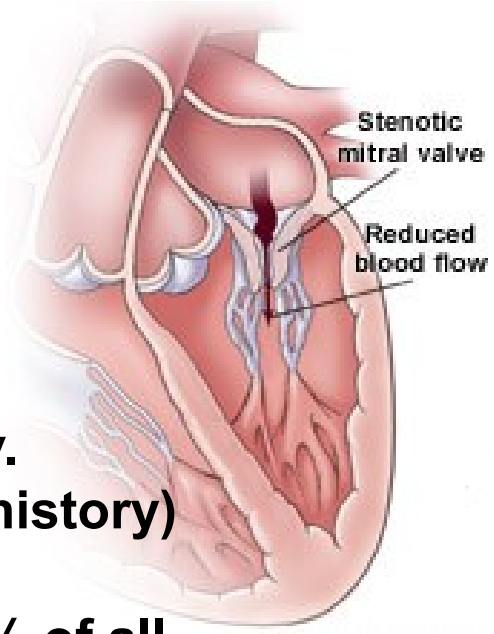

Mi, Tri insufficiency
Ao, Pulm insufficiency
murmur
1
2
3
4
Sounds
systole
diastole
Mi + Tri closure
Ao + Pulm closure


Ao, Pulm stenosis
Mi, Tri stenosis
murmur
1 2 3 4
1 2 3 4
Sounds
systole
diastole
Mi + Tri closure
Ao + Pulm closure

The intensity of murmur (1/6 … 6/6) is not related to hemodynamic severity of valve disease



Mitral stenosis



Primarily the result of rheumatic fever

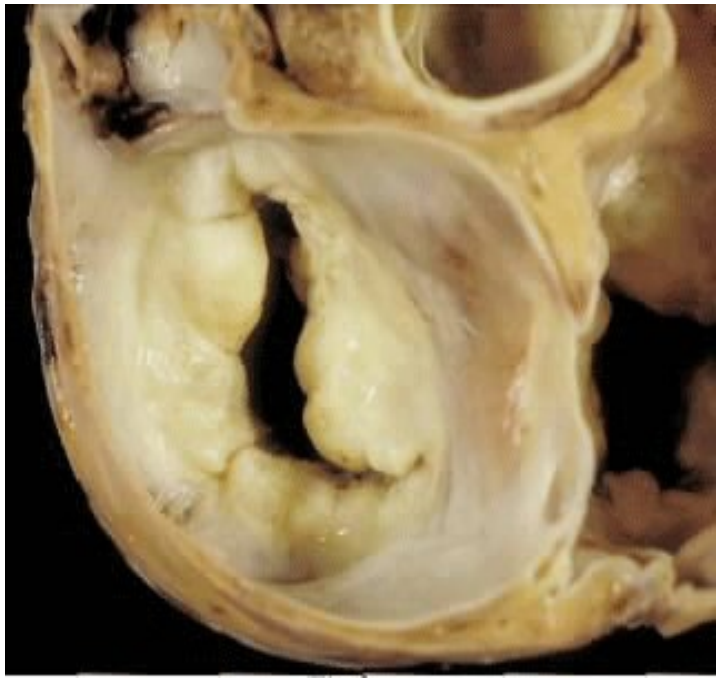
The most frequent post-rheumatic disease (recidiv. tonsilitis, 60% patients with rheumatic fever in history)

Pure or predominant mitral stenosis occurs in 40 % of all patients with rheumatic heart disease

Rarely congenital

Usually combination with mitral insufficiency

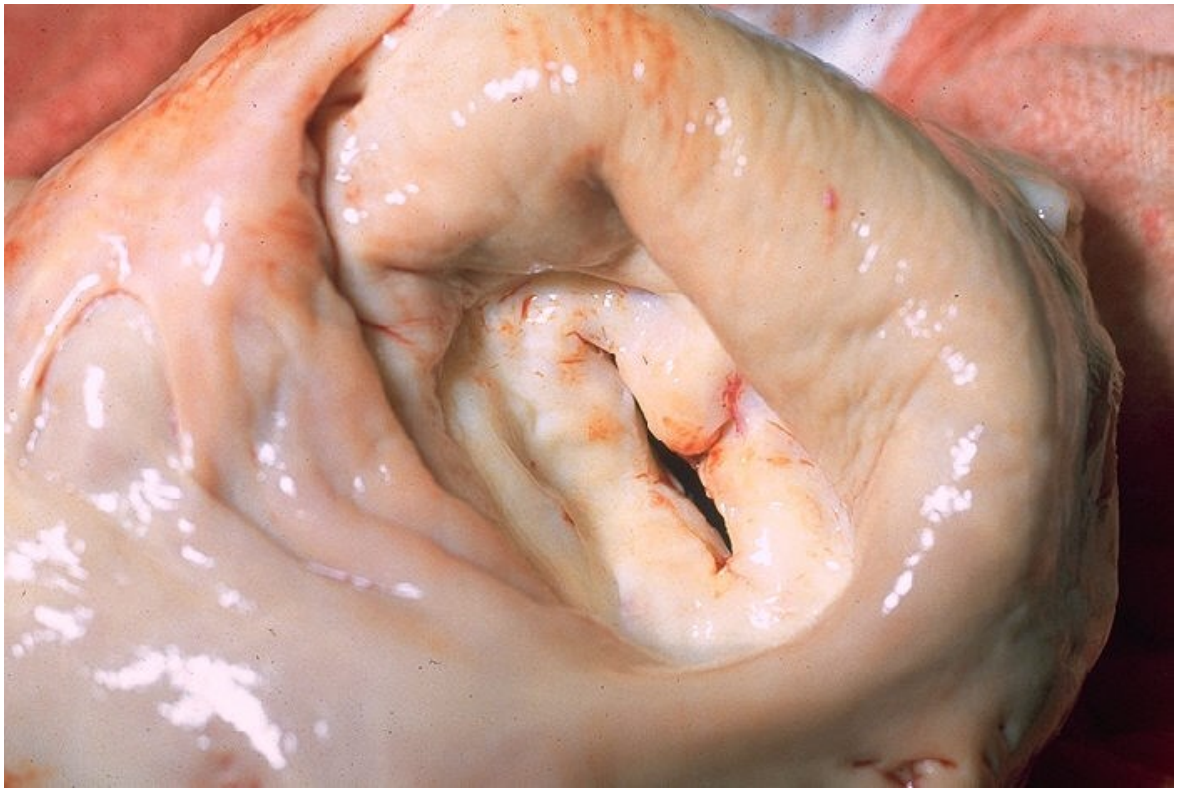
∅ 4-6 cm^2 → 0,5-2 cm^2



Mitral stenosis

Pathogenesis:

♥ **scarring + fusion of valve apparatus**



Mitral stenosis

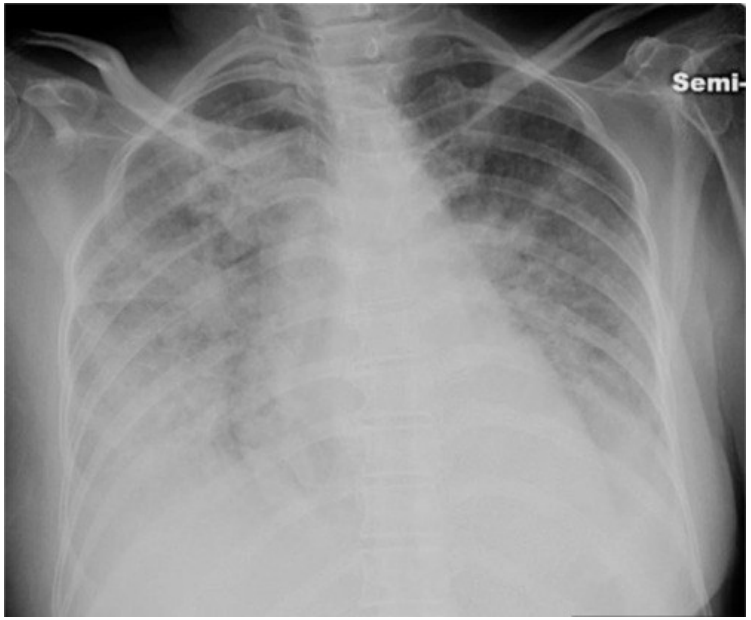
Pathogenesis:

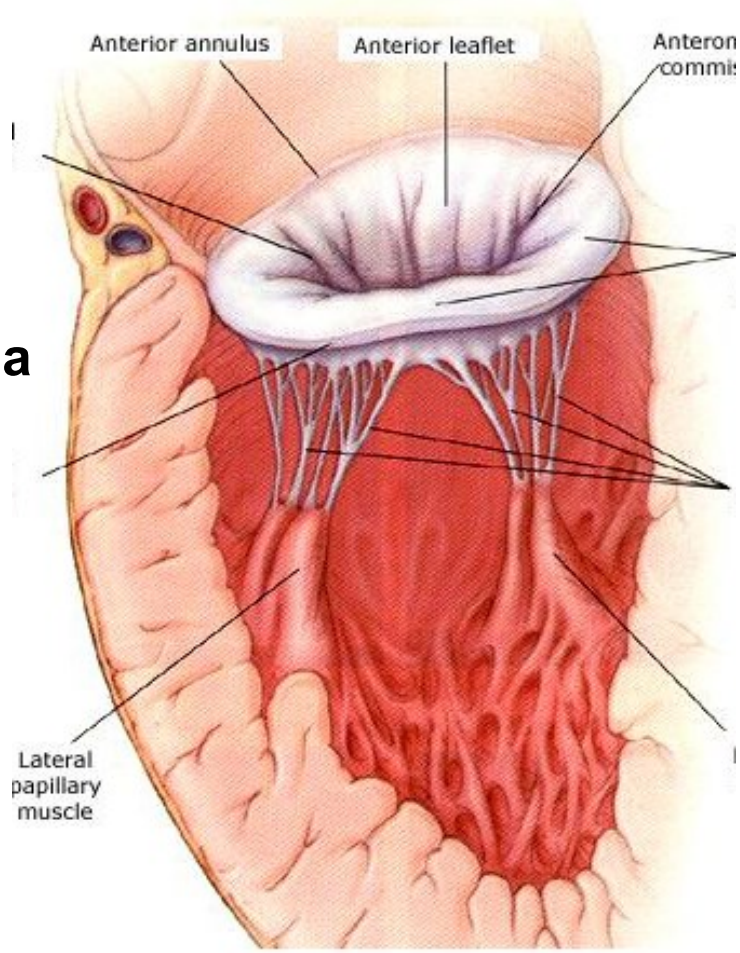
- ♥ scarring + fusion of valve apparatus
- ♥ retardation of quick phase of LV filling (+ participation of atrial fibrillation)
  → ↓CO, poor tolerance of exercise
- ♥ LV hypertrophy / dilation
  → arrhythmia
  → thrombosis / embolism
- ♥ postcapillary pulmonary hypertension
  → pulmonary edema / pulmonary vasoconstriction
  → „tricuspidalisation“ (RV dilation / hypertrophy)

Mitral stenosis

Symptoms:

- ∅ 4 - 6 cm^2 ... normal valve area
- ∅ 1,5 - 2,5 cm^2 ... minimal symptoms
- ∅ 1 - 1,5 cm^2 ... no symptoms at rest
- ∅ ≤ 1 cm^2 … severe stenosis, pulm. edema



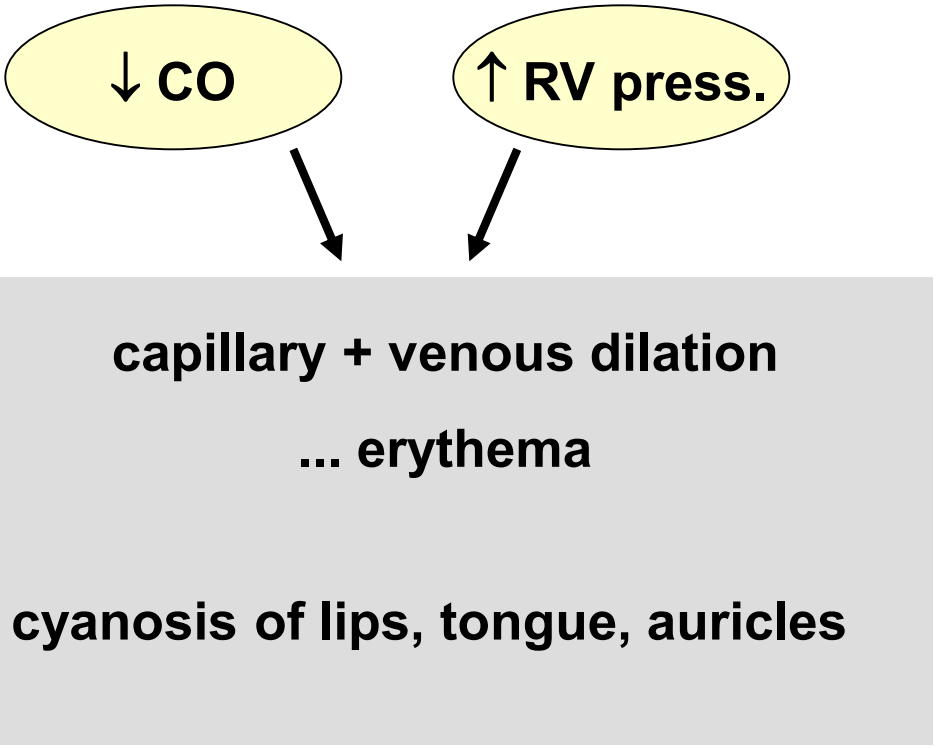


Mitral stenosis

Symptoms:

- **fatigue, poor tolerance of exercise**
- **palpitations**
- **cough**
- **left sided failure (orthopnea)**

Mitral stenosis

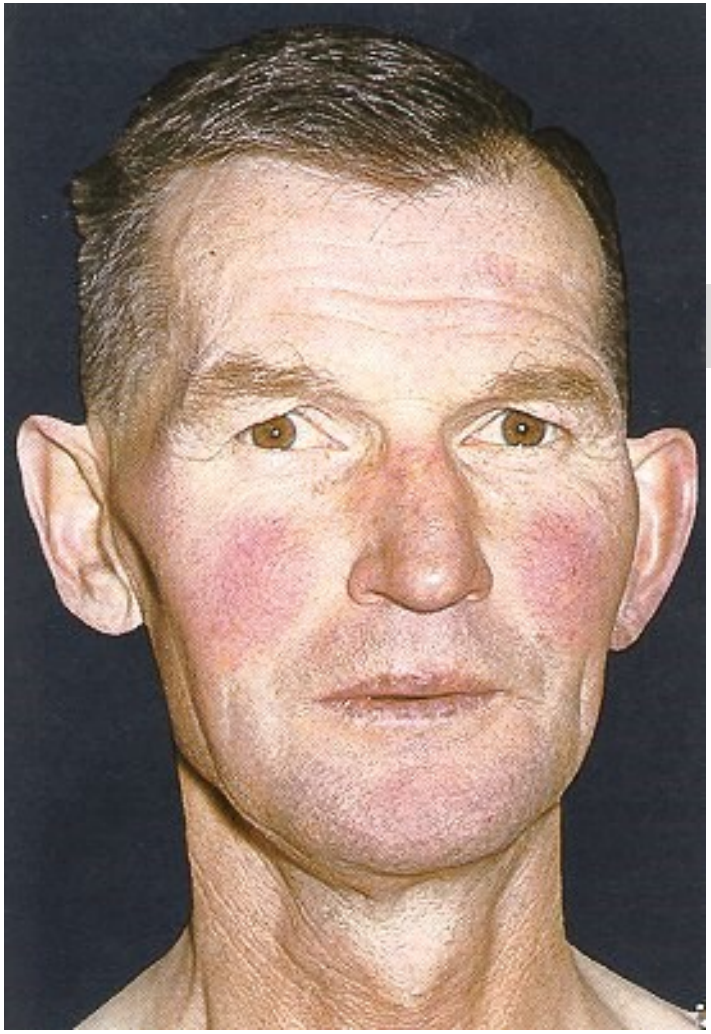


Mitral stenosis

facies mitralis

Flush and small venectasies are in contrast to pale surrounding, lip cyanosis. Dyspnoe, dry cough. Palpitation in atrial fibrillation.

Mitral stenosis

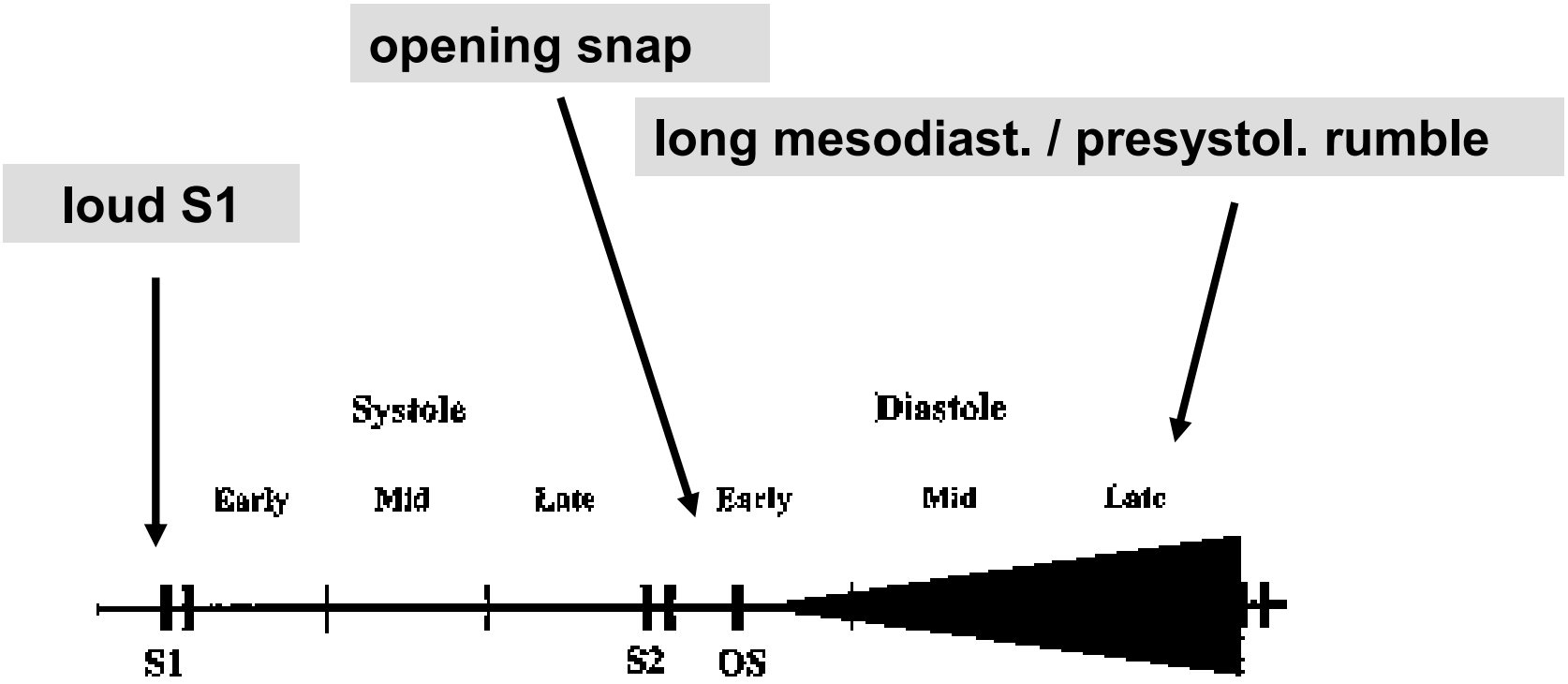


facies mitralis

Flush and small venectasies are in contrast to pale surrounding, lip cyanosis. Dyspnoe, dry cough. Palpitation in atrial fibrillation.

Mitral stenosis

Diagnostics: Auscultation / phonocardiography



Mitral stenosis

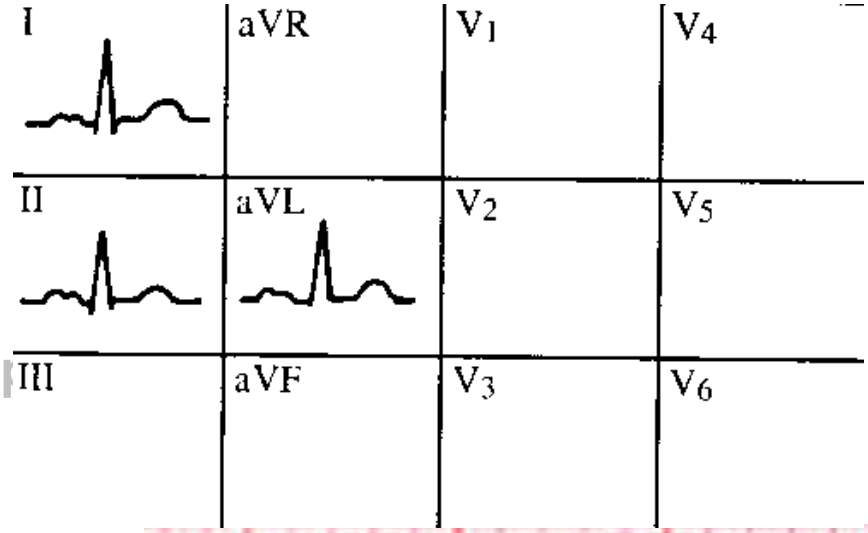
Diagnostics:

Phonocardiography

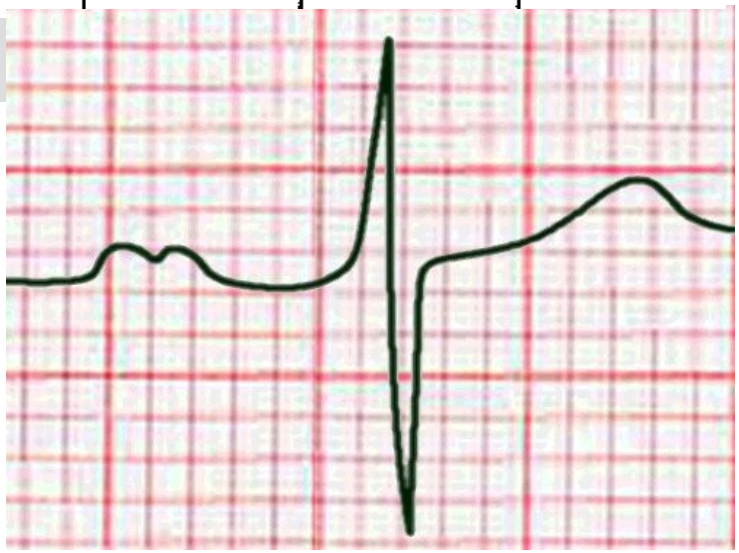
ECG

Chest X-ray – LV enlargement, p

Echocardiography (!)



P mitrale



wide and biphasic P wave; often atrial fibrillation

Mitral stenosis

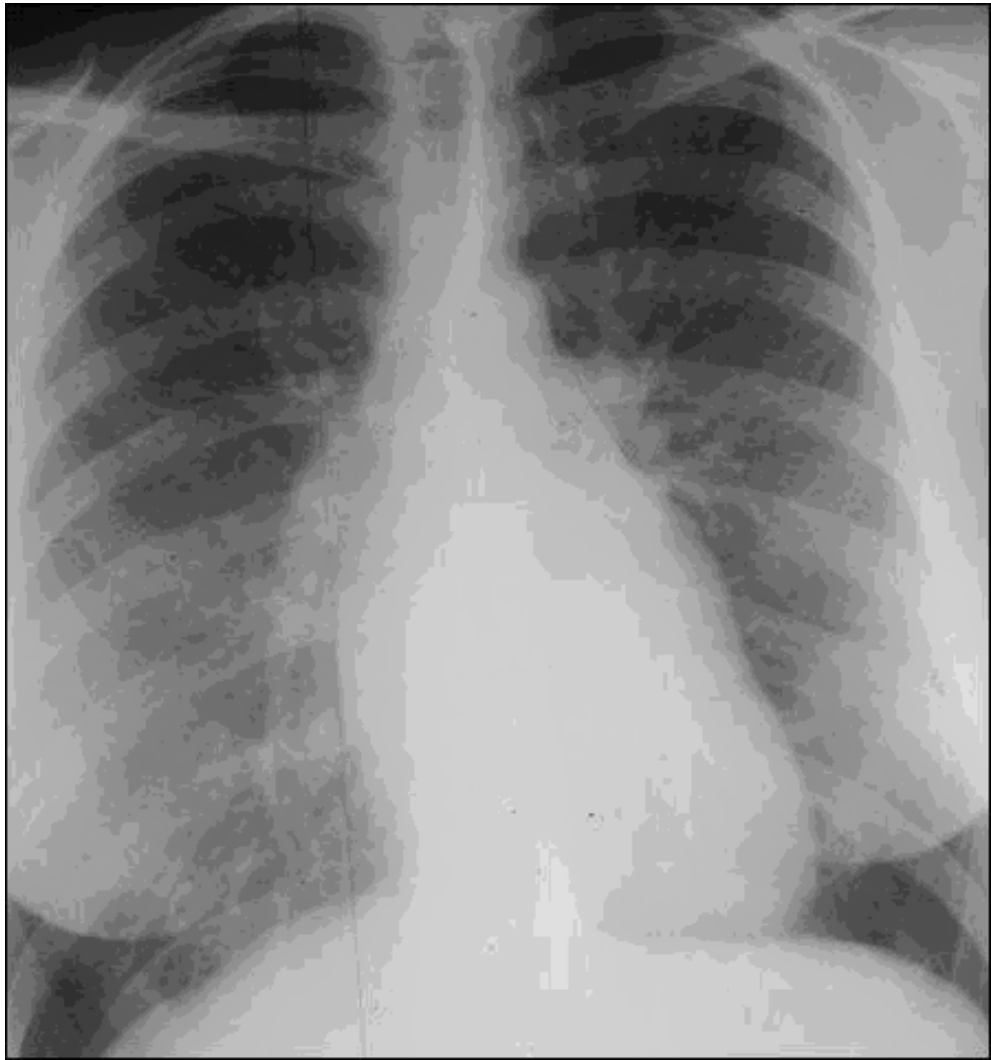
Diagnostics:

Phonocardiography

ECG

Chest X-ray

Echocardiography (!)



LV enlargement

prominent pulmonary veins

Mitral stenosis

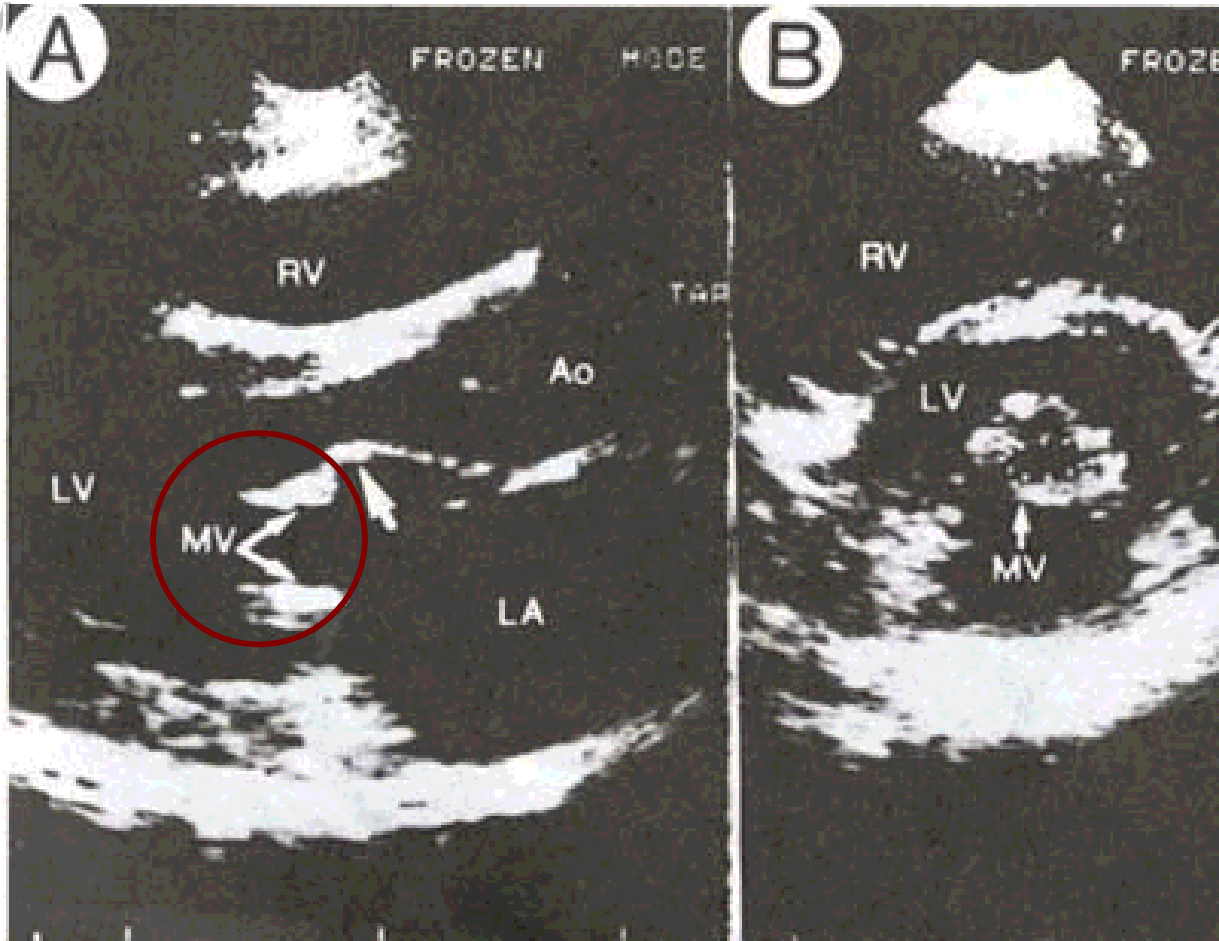
Diagnostics:

Phonocardiography

ECG

Chest X-ray

Echocardiography



Mitral stenosis

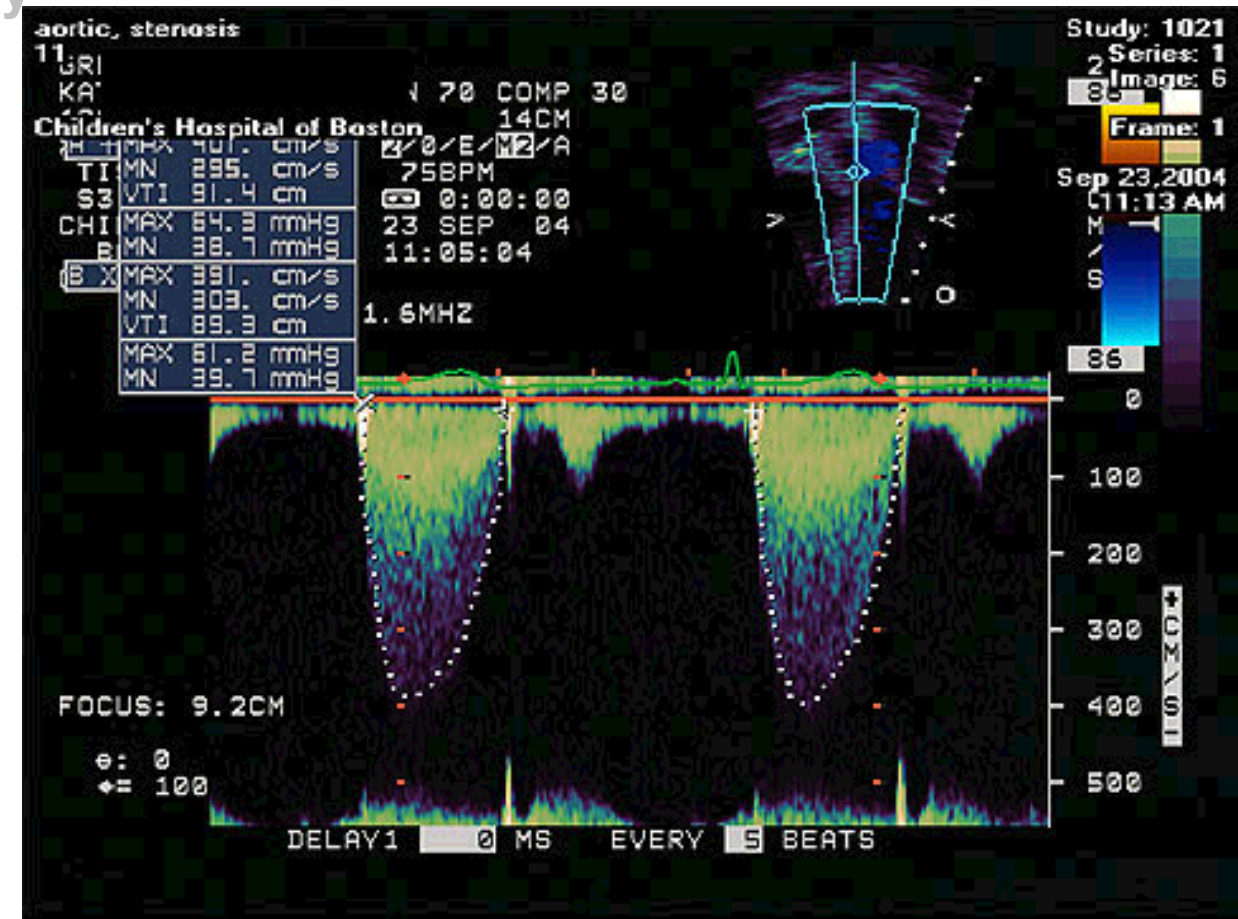
Diagnostics:

Phonocardiography

ECG

Chest X-ray

Echocardiography



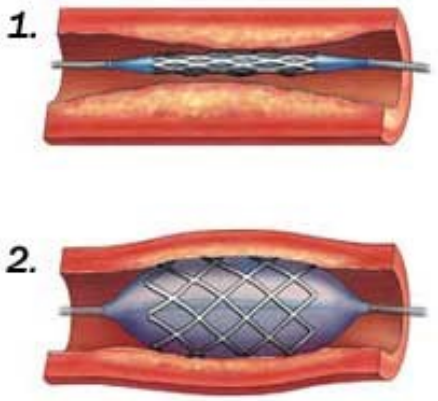
Mitral stenosis

Complications

- atrial fibrillation
- embolism
- infectious endocarditis
- recidiv. bronchitis

Therapy

- catheterization (balloon valvuloplasty);
- surgical replacement of mitral valve



Mitral insufficiency

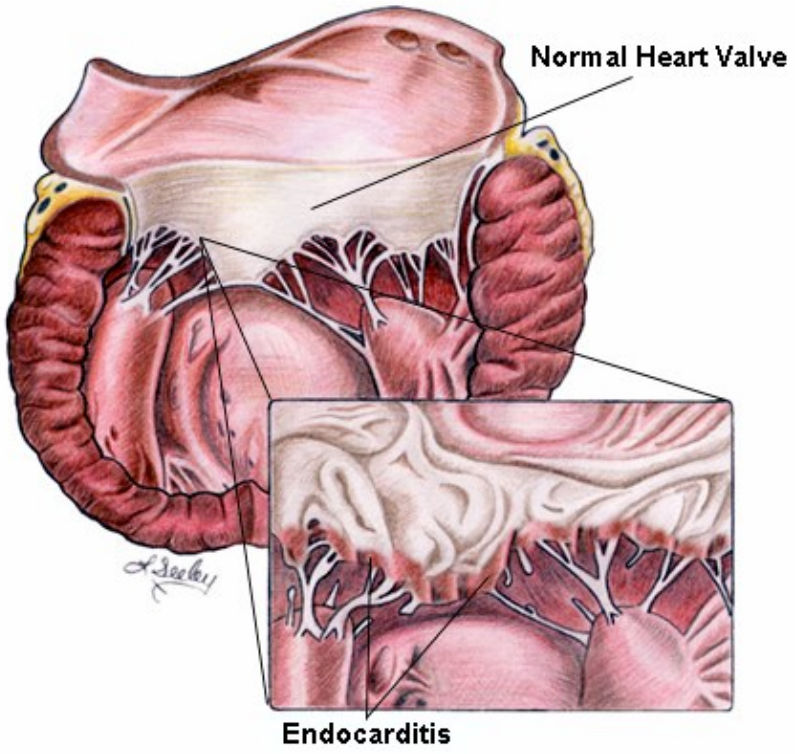
Etiology:

- **mitral valve prolapse**

Mitral insufficiency

Etiology:

- **mitral valve prolapse**
- **rheumatic fever - endocarditis (shortening, thickening of valve) … without latention**



Mitral insufficiency

Etiology:

- **mitral valve prolapse**
- **rheumatic fever - endocarditis (shortening, thickening of valve) … without latention**
- **ischemic disease (dysfunction of papillary muscles)**
- **bacter. endocarditis (valvular rupture, perforation)**
- **obstructive cardiomyopathy**
- **“relative insufficiency” (LV dilation)**

Mitral insufficiency

Pathogenesis:

♥ **regurgitation of CO from LV to LA (> 50 %)**

→ **LA hypertrophy / dilation (larger than mitr. stenosis)**

→ **decrease of LV systolic filling**

… **long-term good tolerance**

→ **poor tolerance of exercise, dyspnoe, palpitations**

Prognosis:

long latent course

complications (bacterial endocarditis, embolism)

quick terminal progress

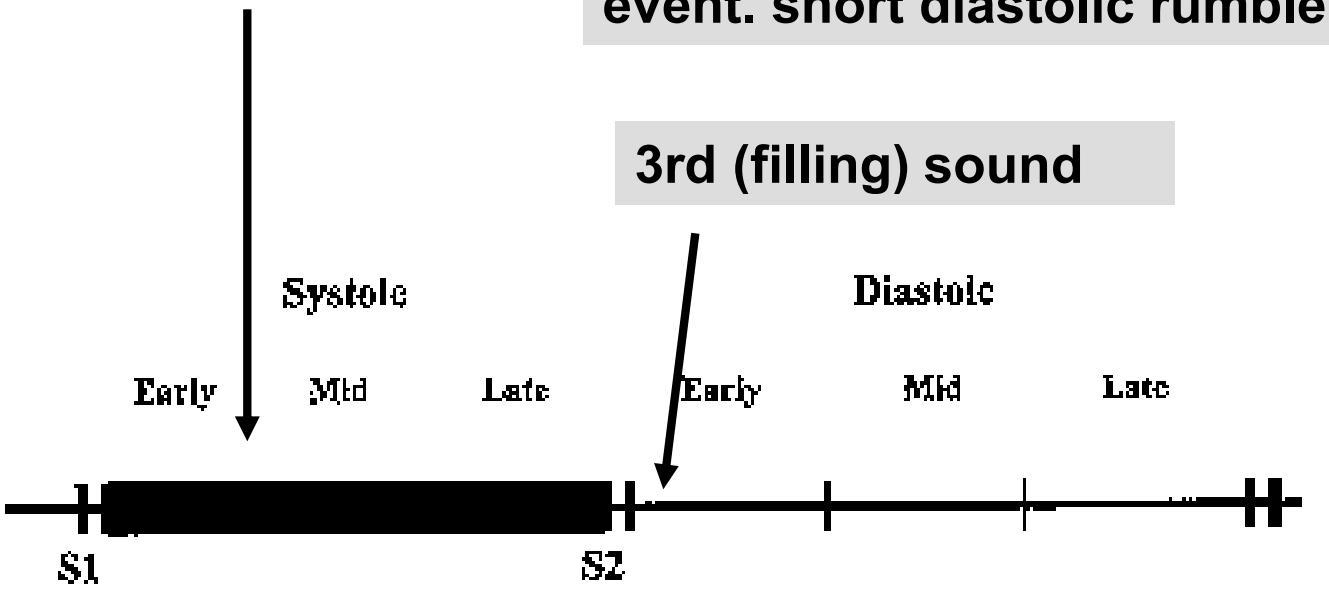
Mitral insufficiency

Auscultation:

holosystolic rumble on apex (systolic whirl)

event. short diastolic rumble

3rd (filling) sound

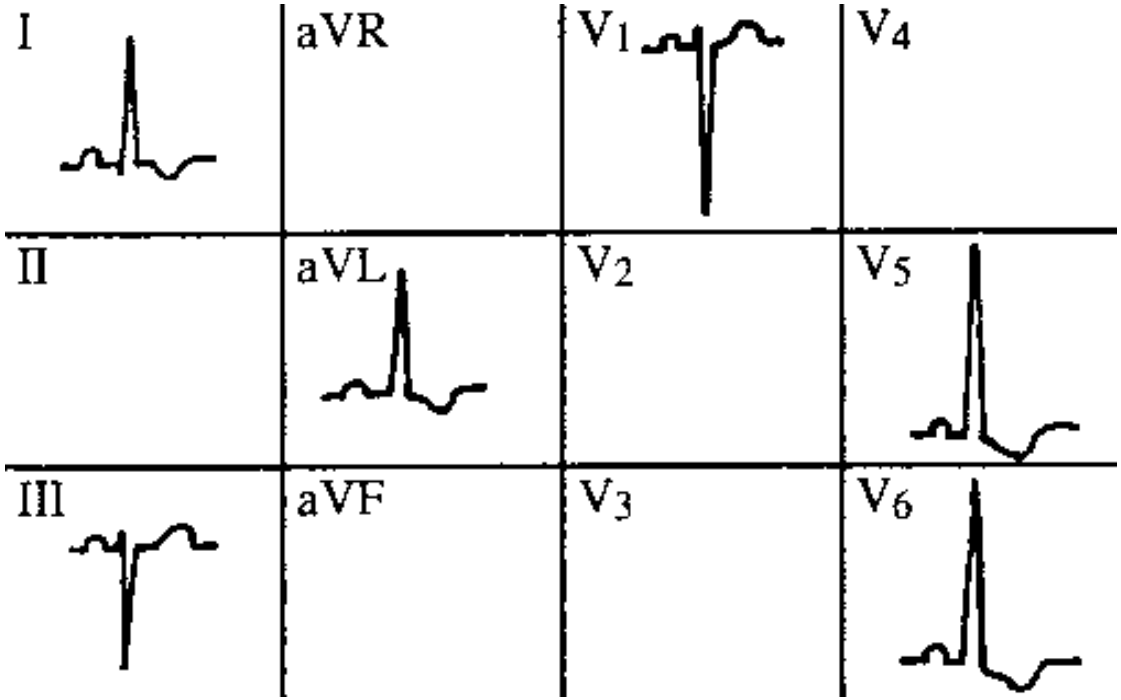


Mitrální insuficience

Diagnostics:

Phonocardiography

ECG – LV hypertrophy or P mitrale, atrial fibrillation



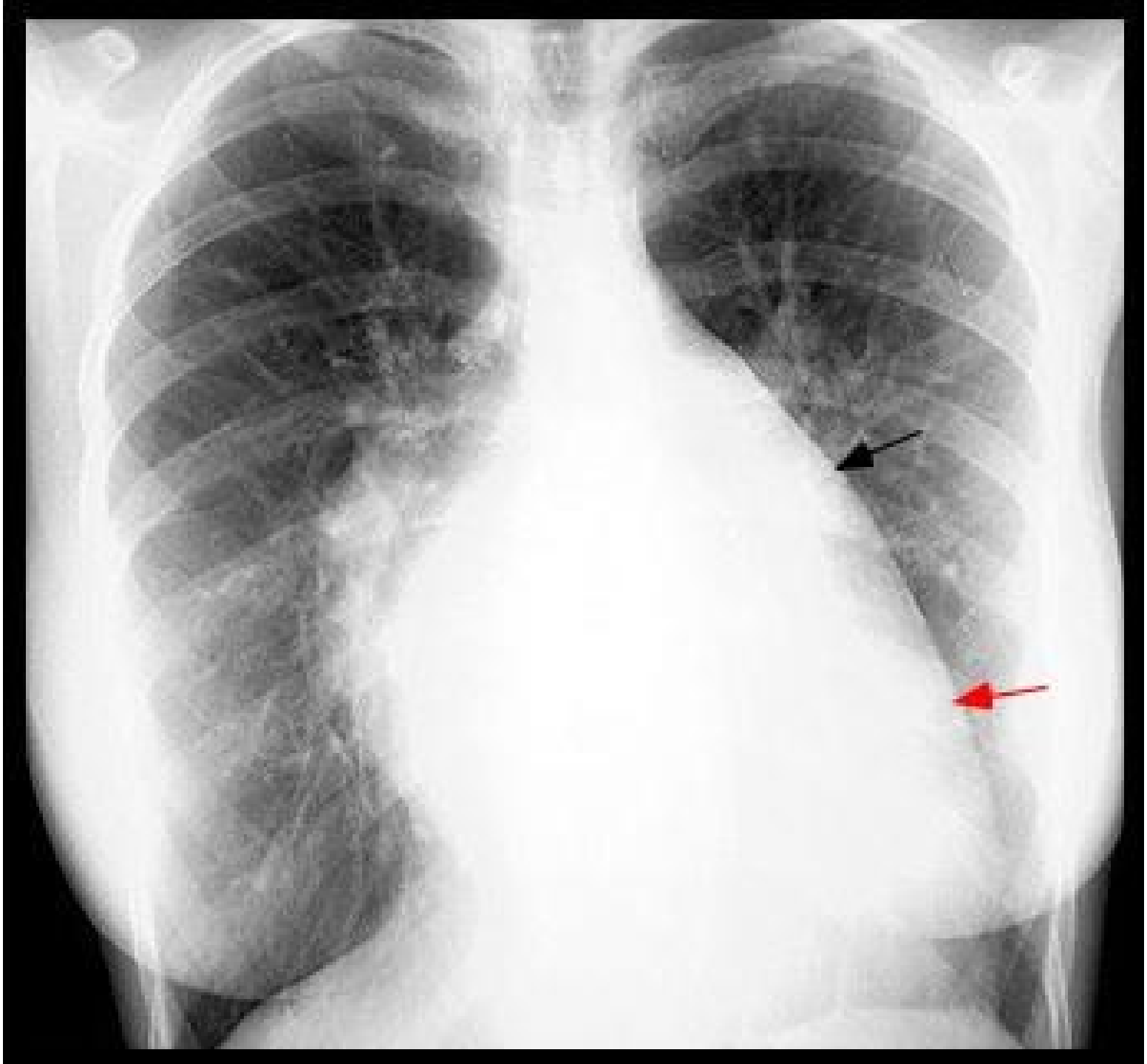
Mitrální insuficience

Diagnostics:

Phonocardiography

ECG

Chest X ray – heart hypertrophy / dilation



Mitrální insuficience

Diagnostics:

Phonocardiography

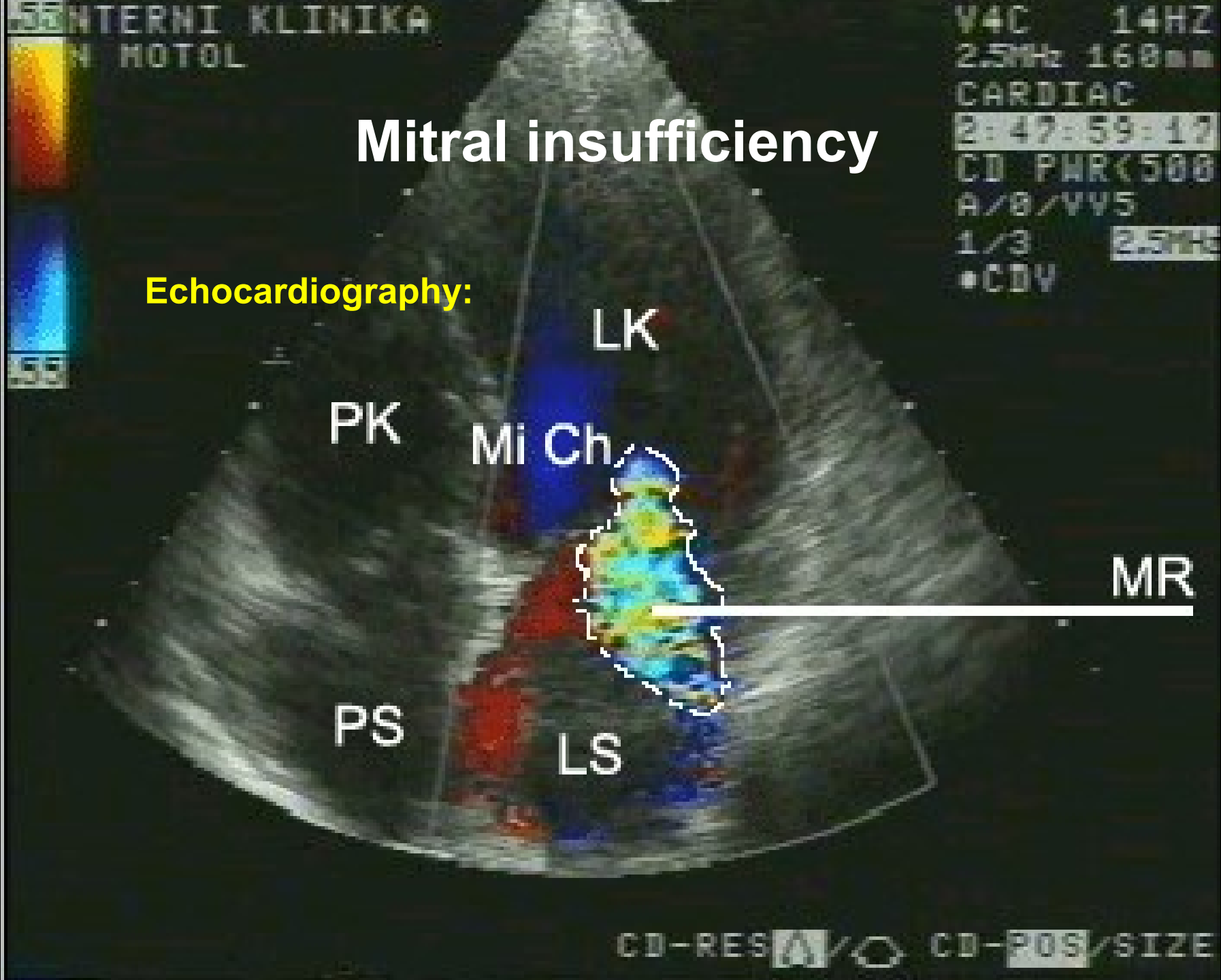
ECG

Chest X ray

Echocardiography (!)

Mitral insufficiency

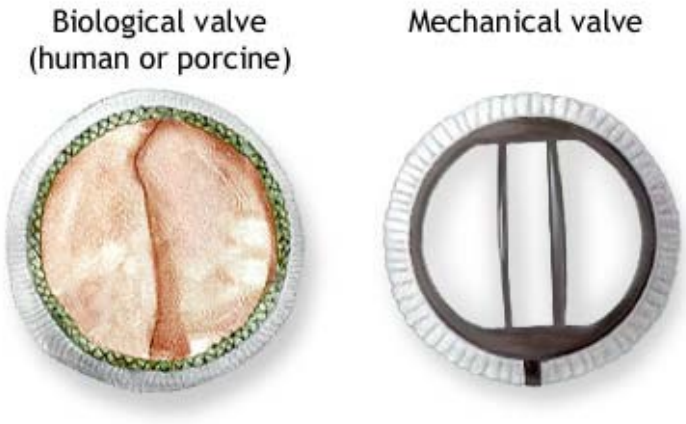
Echocardiography:



Mitral insufficiency

Complications

- atrial fibrillation,
- heart failure, pulmonary edema,
- infectious endocarditis



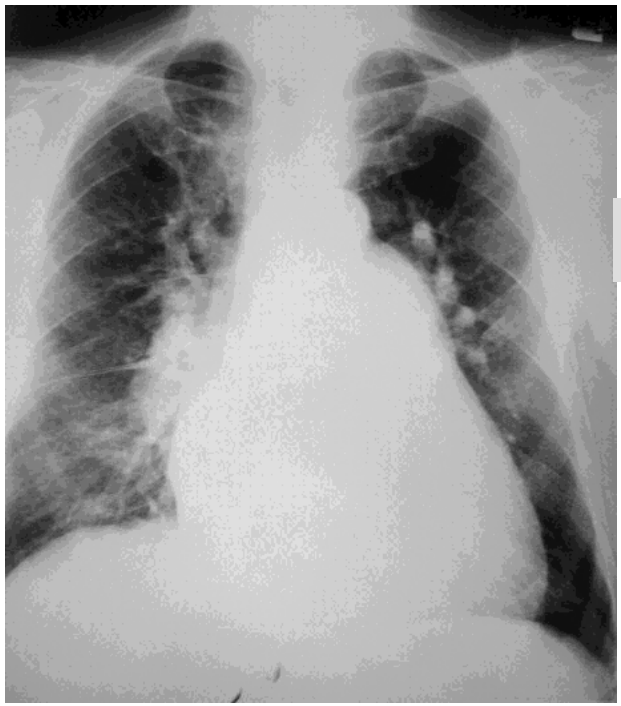
Therapy

- surgical replacement of a valve

Combined mitral disease

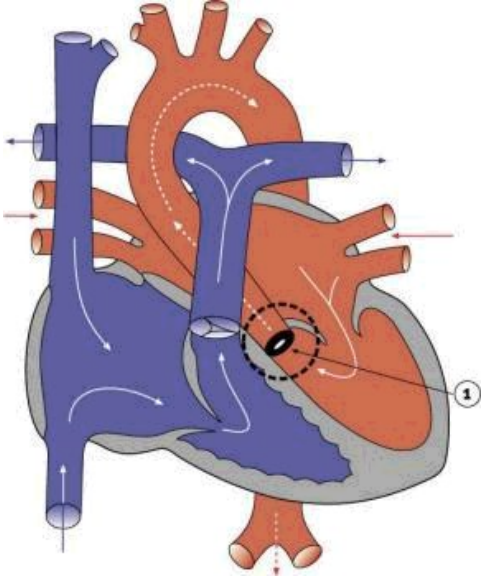
Mitral ostium > 1 cm^2 … dominant insufficiency … LV hypertrophy

Mitral ostium < 0,6 cm^2 … dominant stenosis … RV hypertrophy, pulmonary hypertension

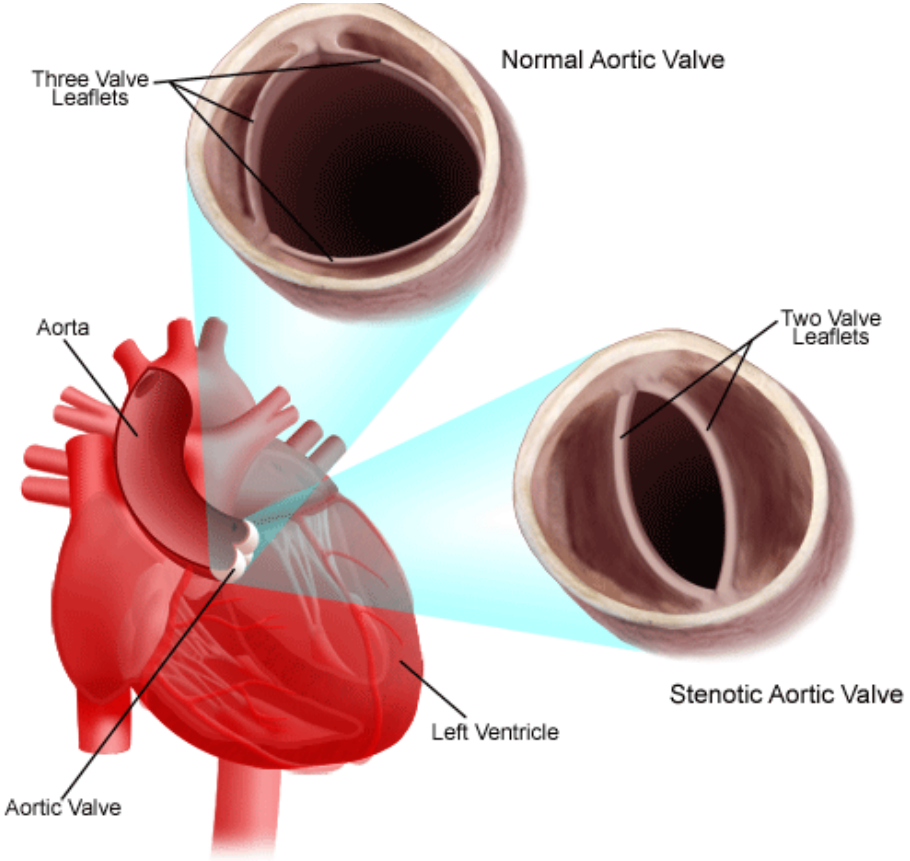


combined mitral disease

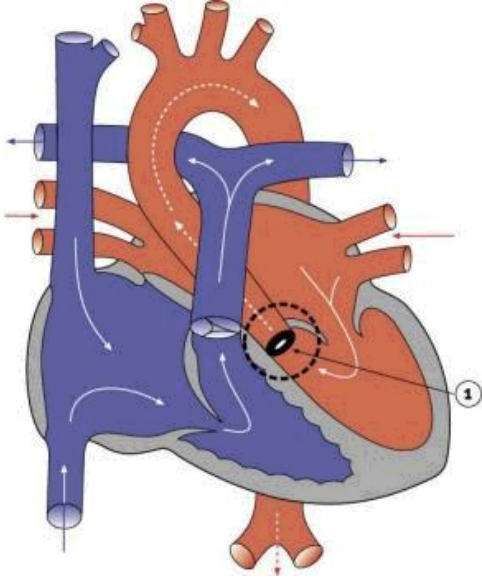
Aortic stenosis



normal size ∅ 3 cm²
hemodyn. important stenosis ... ¼ (= 0,7 cm2)



Aortic stenosis



normal size ∅ 3 cm^2
hemodyn. important stenosis ... ¼ (= 0,7 cm^2)

Etiology:

- **rheumatic fever (combination with insufficiency)**
- **congenital (valvular adhesion)**
- **degenerative (sclerosis) (elderly patients, benign course)**

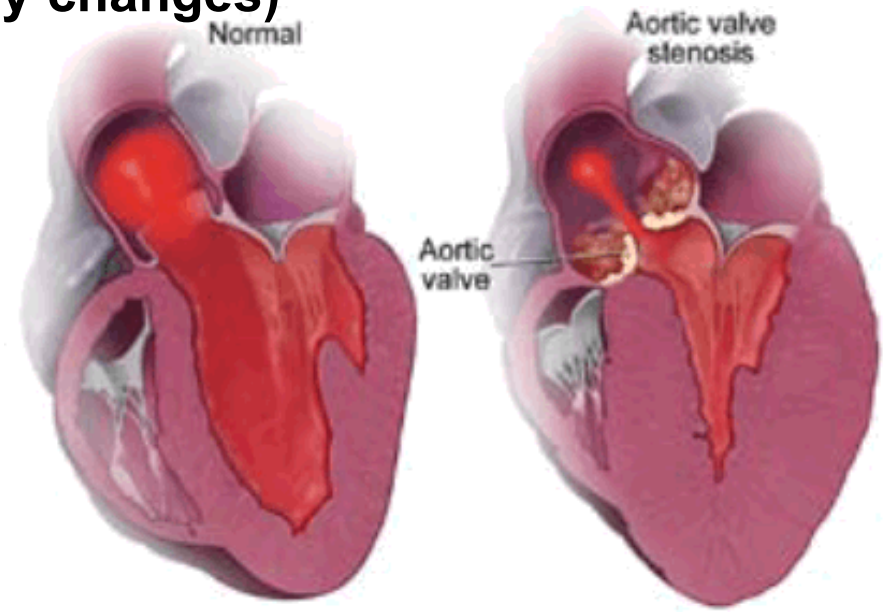
Localization:

- **supravalvular (necking above valve)**
- **valvular**
- **subvalvular**

Aortic stenosis

Pathogenesis:

- ♥ ↑ pressure systolic gradient between LV and the aorta
  - → *pulsus parvus, pulsus tardus*
  - → systolic pressure 100-110 mm Hg
  - → normal CO, but no ↑ during exertion (…syncope, dizziness)
- ♥ LV hypertrophy → LA hypertrophy → pulmonary propagation (concentric HY … without X-ray changes)



Aortis stenosis

Prognosis:

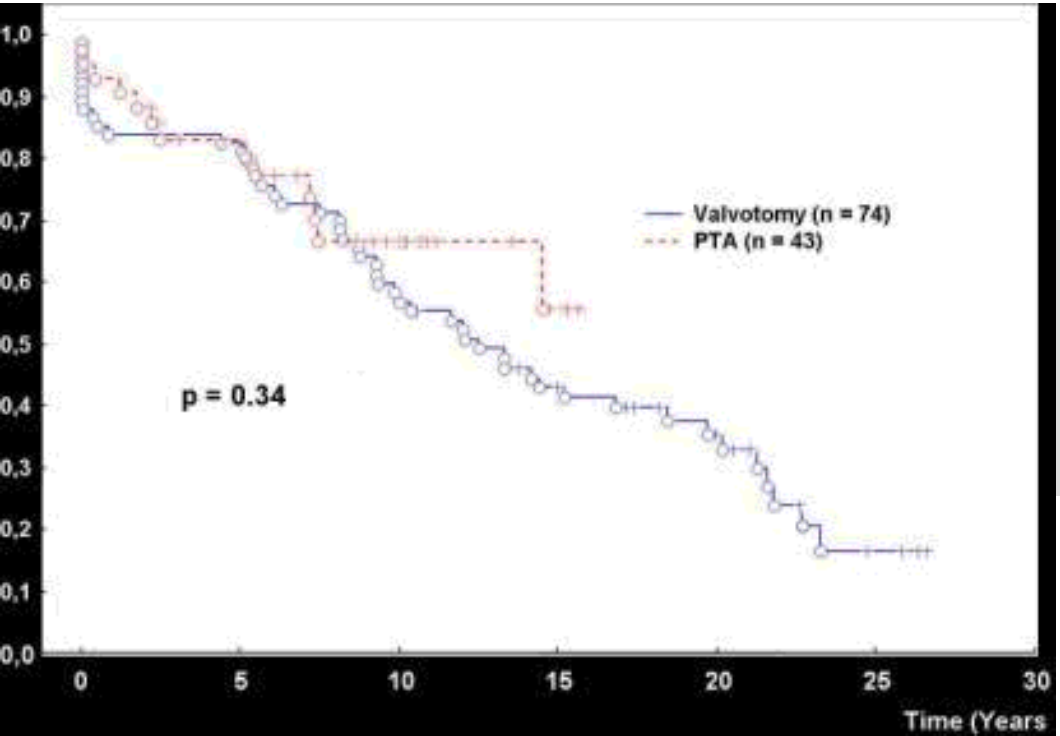
slow progression (...surgery: systolic gradient 50-70 mm Hg)
complications: bacterial endocarditis, AMI
terminal cardiac failure

- **dyspnoe; poor tolerance of physical activity**
- **syncopes in exercise;**
- **angina pectoris;**
- **systolic murmur in 2nd intercostal space in right parasternal line**

Aortis stenosis

Prognosis:

slow progression (...surgery: systolic gradient 50-70 mm Hg)
complications: bacterial endocarditis, AMI
terminal cardiac failure



Survival curve of patients after palliative surgery - valvotomy, or PTA (balloon valvuloplastic)

Aortic stenosis

Diagnostics:

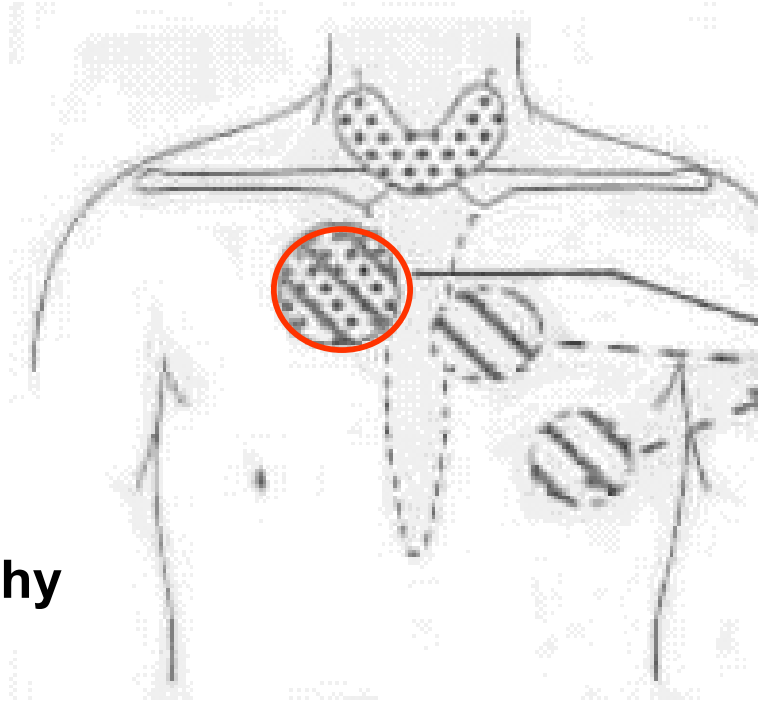
Phonocardiography

ECG – LV hypertrophy

Chest X-Ray

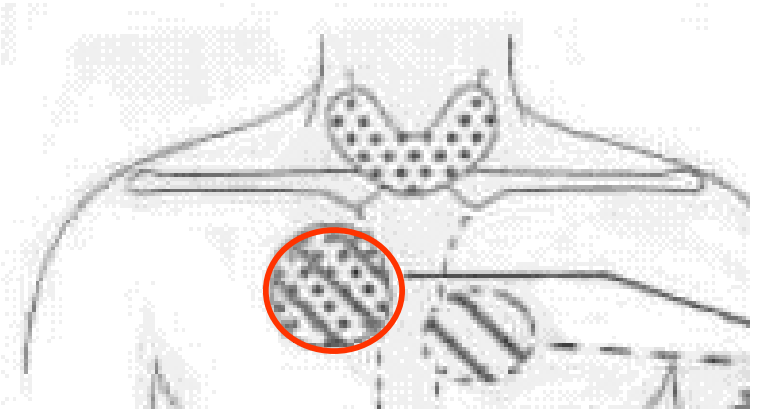
Echocardiography

Heart catheterization - ventriculography



Aortis stenosis

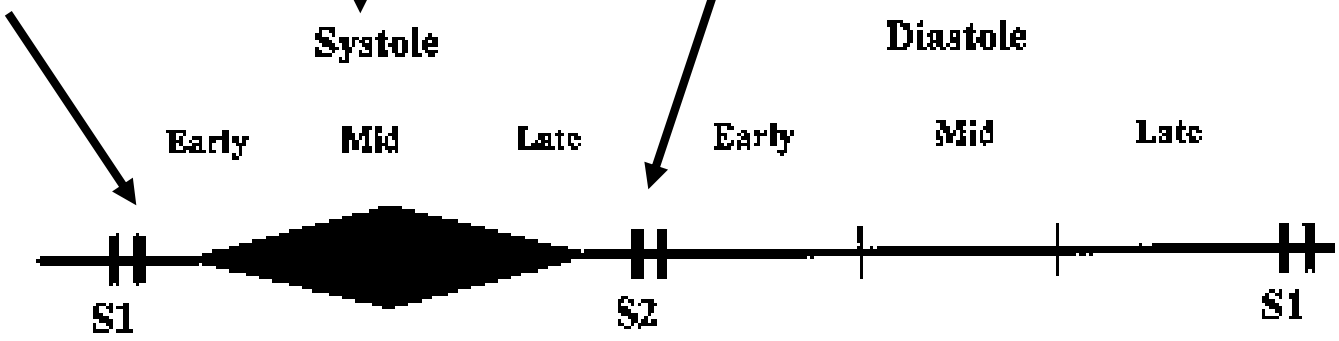
Auscultation:



eject systolic murmur

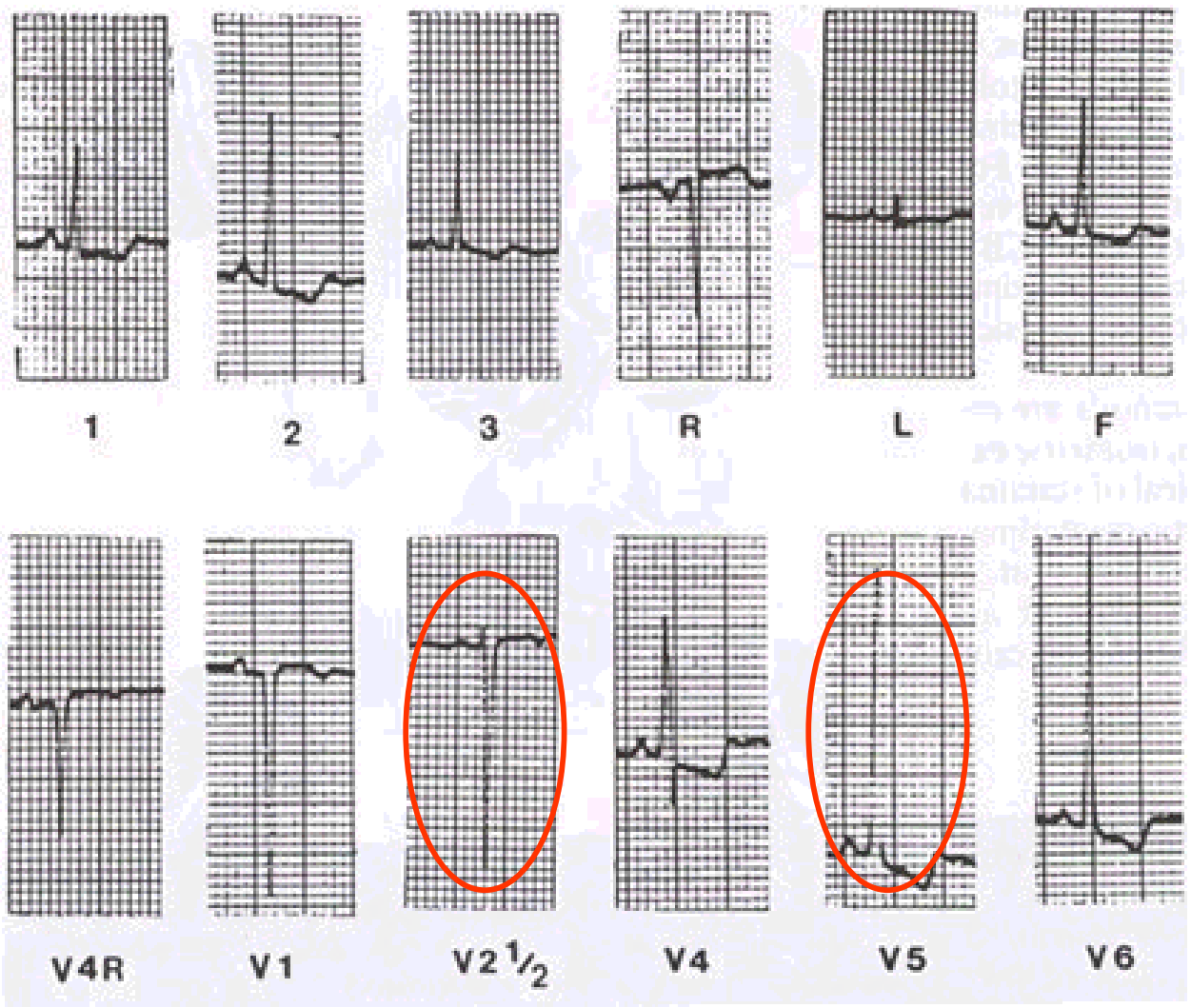
early systolic click

S2 cleavage in exspirium



Aortis stenosis

ECG: LV overloading



Aortis stenosis

Cardio-CT:

Diffuse calcification of all 3 aortic commisuras, mild stenosis in 66-yr. male

Aortic stenosis

Complications

- infectious endocarditis,
- cardiac failure
- sudden death

Therapy

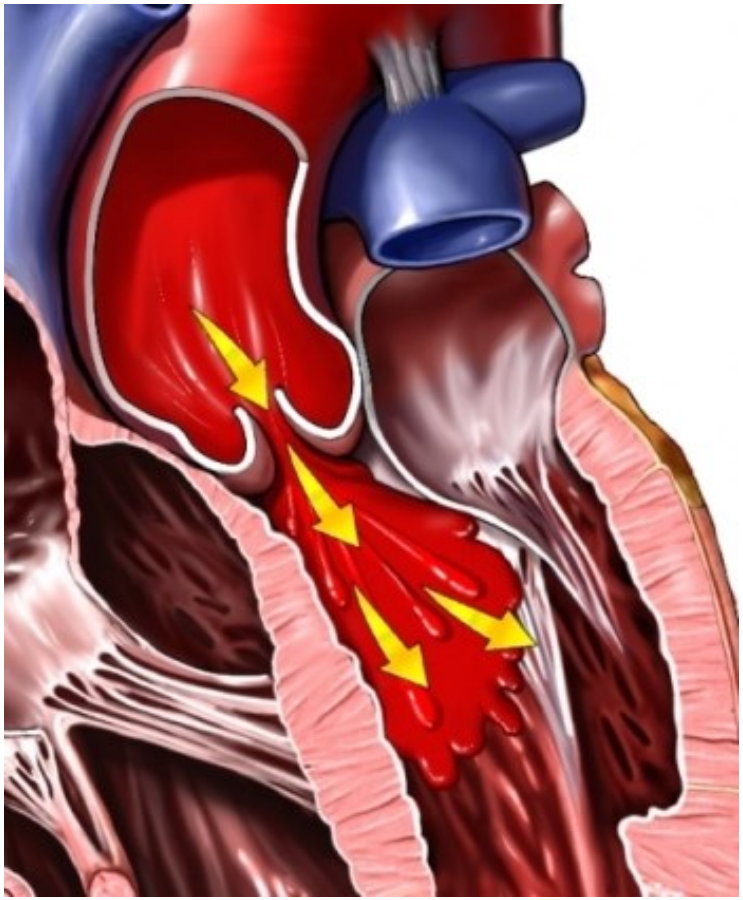
- surgical valve replacement

Aortic valve replacement

Aortic insufficiency

Etiology:

- **rheumatic fever - endocarditis !! (without delay, often combined with mitral disease)**
- **bacterial endocarditis**
- **congenital disease**
- **dissection of aortic aneurysm**
- **pox**
- **“relative insufficiency” = dilation of aorta (e.g. during pox)**



Aortic insufficiency

Pathogenesis:

depends on ∅ valve, gradient, and LV compliance

- **(compensatory) LV hypertrophy / dilation**
- **(compensatory) ↓ peripheral resistance**
  - **→ Corrigan's pulse (high, quick, ↑ pressure amplitude)**
  - **→ Quincke capillary pulsation**
- **good exertion tolerance (due to ↓ vascular resistance)**

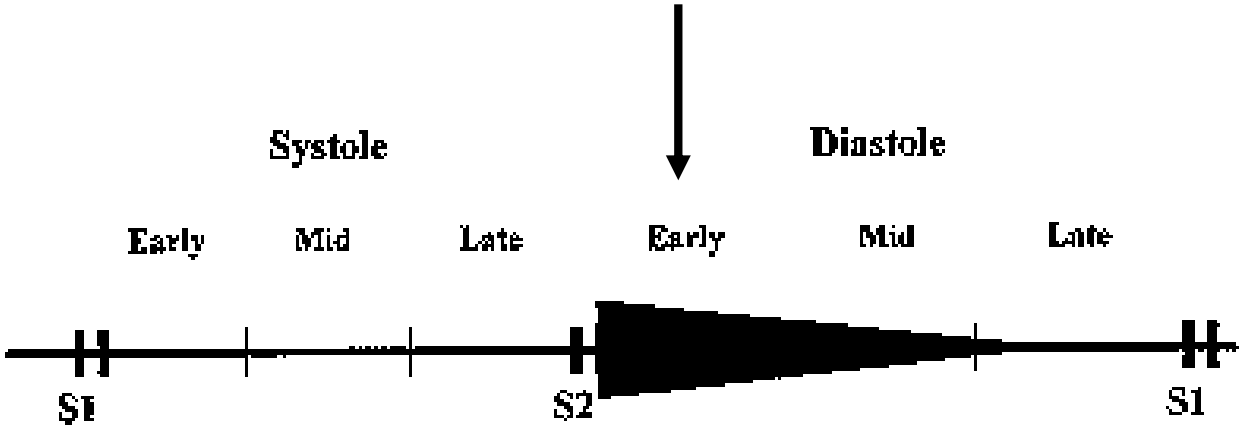
Prognosis:

10x years latent course, palpitations
complications: bacterial endocarditis
terminal cardiac failure

Aortic insufficiency

Auscultation:

decrescend diastolic murmur



Aortic insufficiency

Diagnostics:

echo,
heart catheterization (ventriculography)

Prognosis:

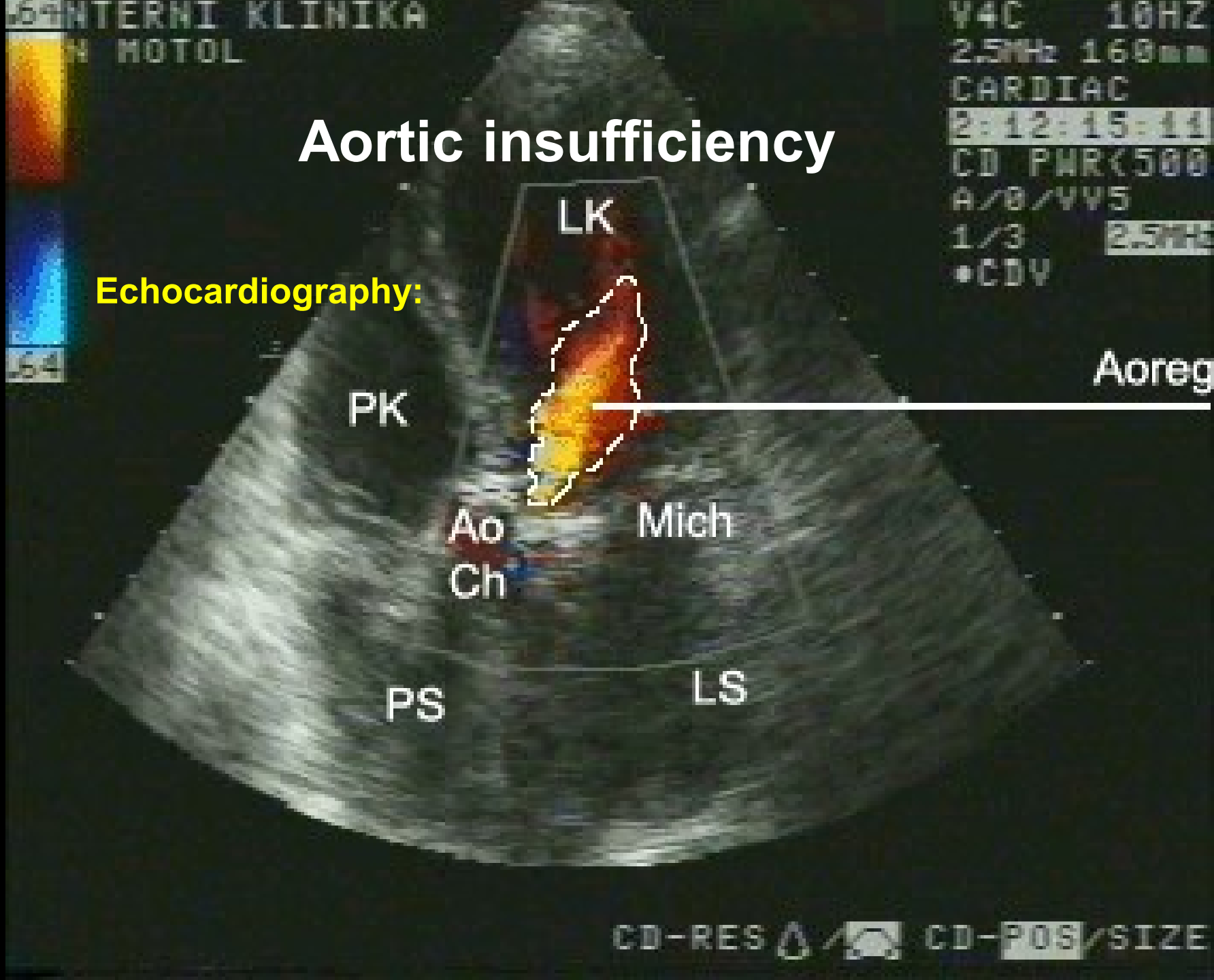
10 x years latent course, palpitations
complications: bacterial endocarditis
terminal cardiac failure

Therapy:

valve replacement

Aortic insufficiency

Echocardiography:



Congenital heart defects

Septal defects and ductus arteriosus

Atrial / ventricular septal defect,

Persistent truncus arteriosus

Obstruction defects

Coarctation of the aorta

Dislocation defects

Transposition of the great vessels

Ebstein anomaly

Combined defects

Fallot tetralogy

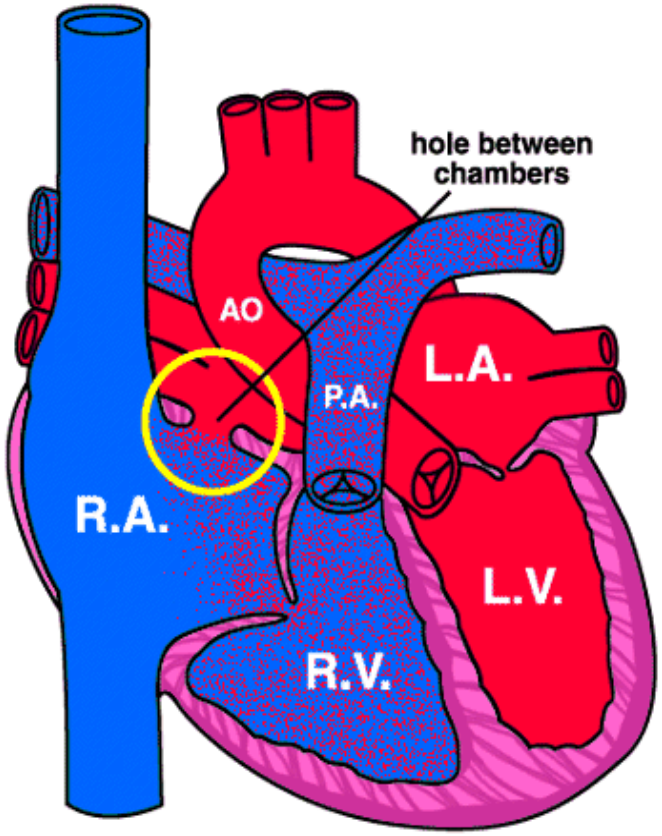
Eisenmenger syndrome

Congenital heart defects

Incidence: 9 / 1000 newborns

Atrial septal defect

- Shunt heart defect
- Unclosed foramen ovale
- Oxygenated blood flow from left atrium to right atrium



Atrial septal defect

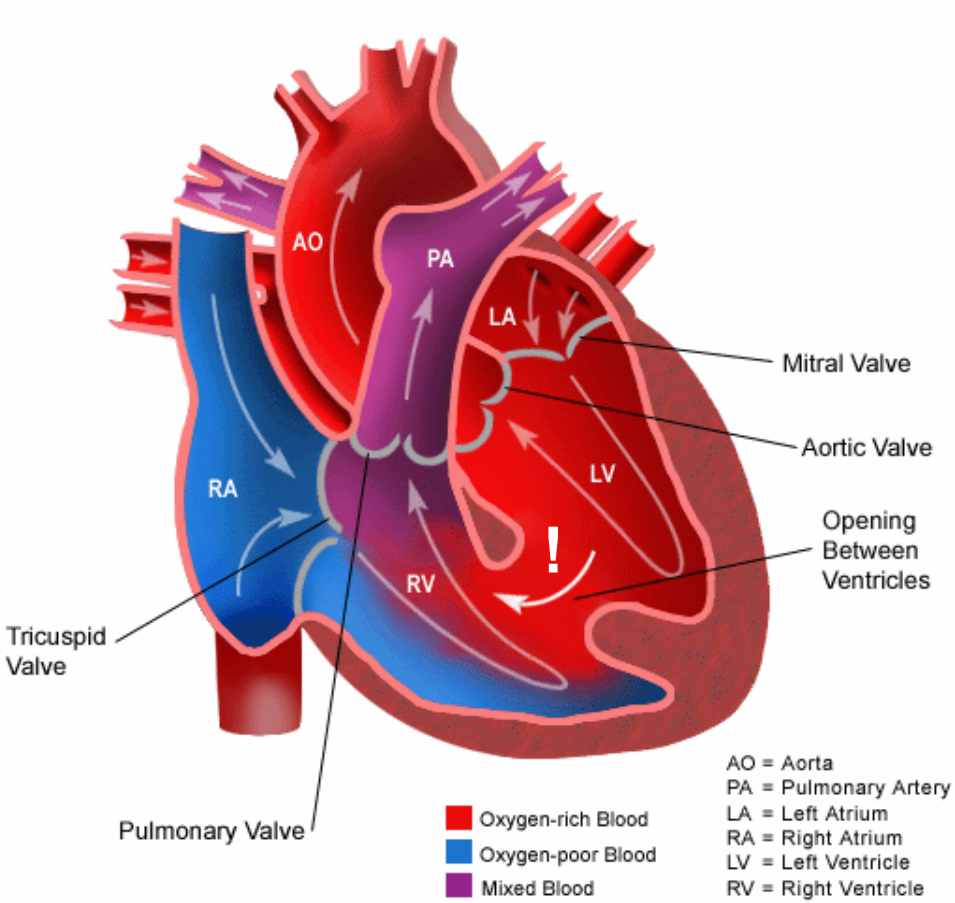
- Shunt heart defect
- Unclosed foramen ovale
- Oxygenated blood flow from left atrium to right atrium

- **Minimal defect (in 30% adults) – asymptomatic**
- **Small defect - dg. at age 30 - 40 – dyspnoe, fatigue, arrythmia**
- **Large defect – dg. in newborn, chronic heart failure**

- **Diagnostics: ECG, Chest X-Ray, ECHO**
- **Therapy: catheterization or cardiac surgery**

Ventricular septal defect

- **Unclosed septum between LV and RV**
- **LV → RV blood flow**



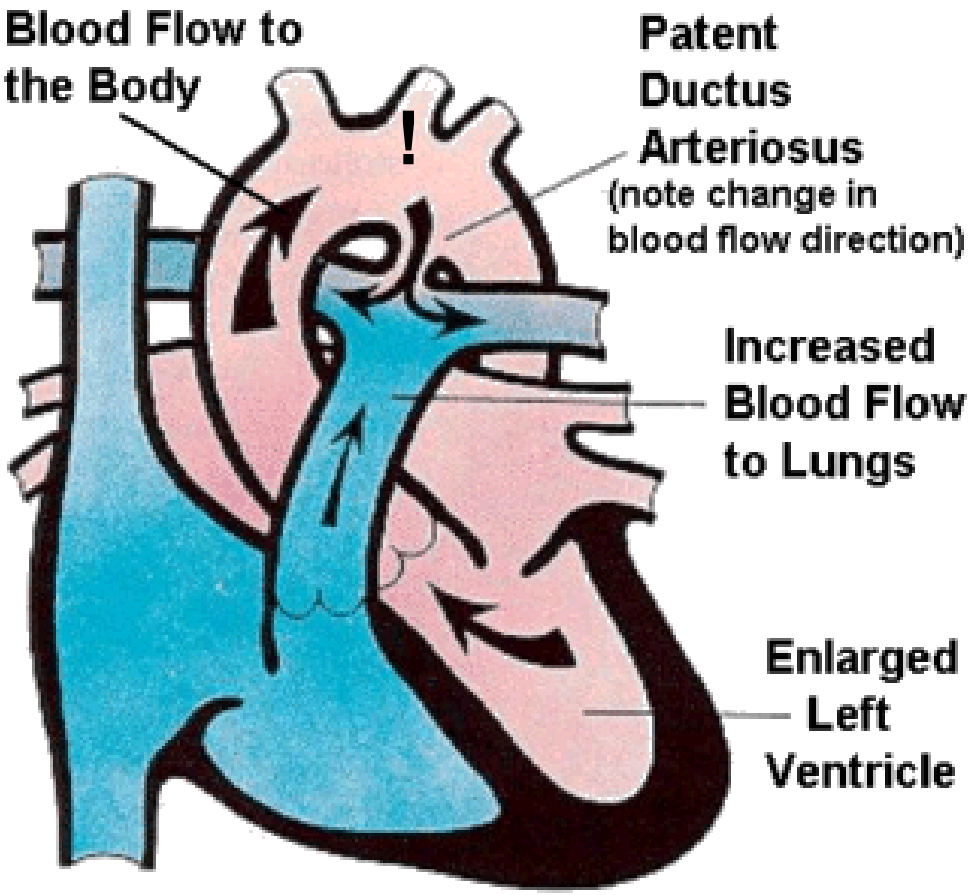
Ventricular septal defect

- Unclosed septum between LV and RV
- LV $\rightarrow$ RV blood flow

- **Small defect**: asymptomatic, loud systolic murmur in the left parasternal line down
- **Large defect**: cardiac failure in newborns
- Diagnostics: ECG, Chest X-Ray, ECHO
- Therapy: cardiac surgery

Persistent truncus arteriosus

- **Truncus arteriosus = In fetus, a communication between pulmonary artery and aorta (blood flow from pulmonary artery to aorta) – closed shortly after delivery**

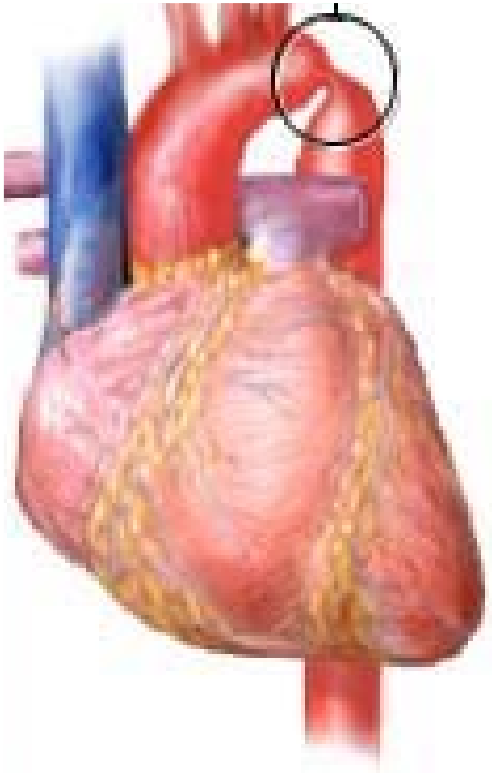


Persistent truncus arteriosus

- **Asymptomatic** at first – due to a high pressure in pulmonary circulation. With a decrease in this pressure, the development of **left-to-right shunt**
- 6-8 weeks after delivery – **intensive continual murmur** (systolic-diastolic) in the left 2nd intercostal space
- Diagnostics: echo
- Large shunt: cardiac failure after delivery
- **Complications**: infectious endocarditis, cardiac failure in adilt age
- **Therapy**: Indomethacin (inhibitor of PG synthesis … vasocon-striction … shunt closure), otherwise surgical treatment

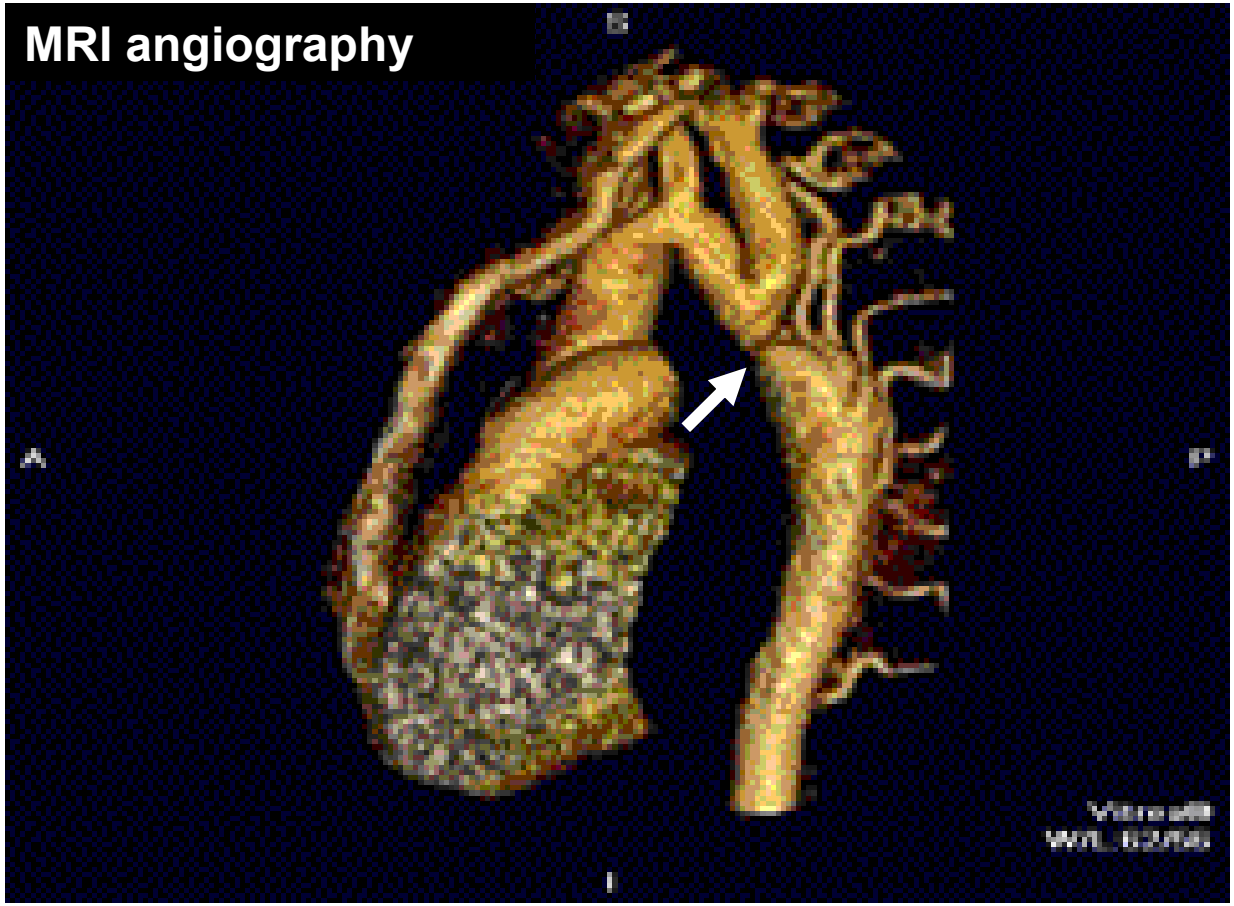
Coarctation of the aorta

- **Obstruction defect**
- **Stenosis of the aorta below a. subclavia sin.**



Coarctation of the aorta

- **Obstruction defect**
- **Stenosis of the aorta below a. subclavia sin.**



Coarctation of the aorta

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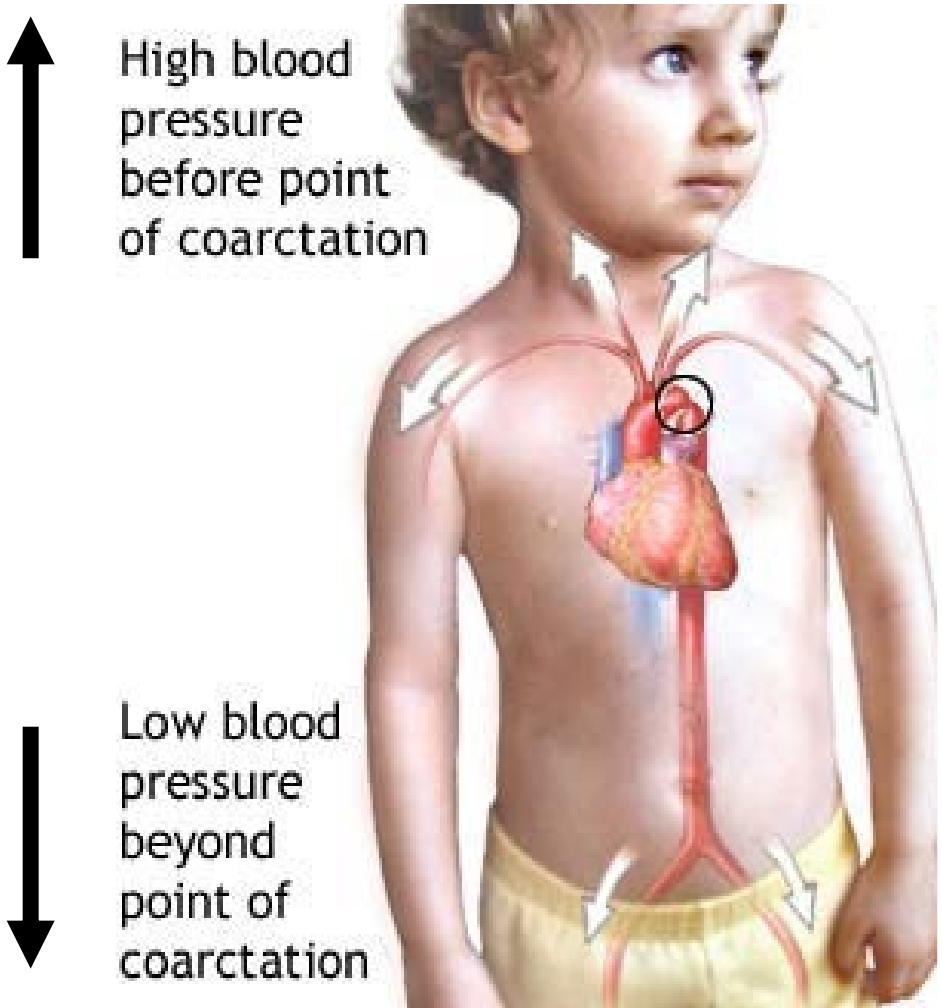
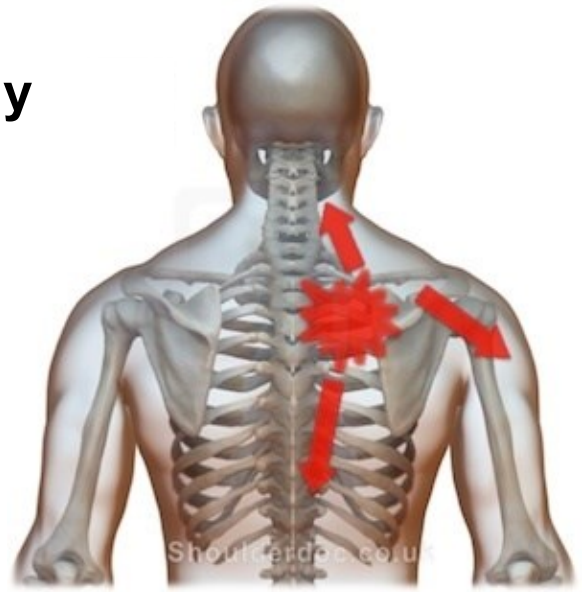


Coarctation of the aorta

- Obstruction defect
- Stenosis of the aorta below a. subclavia sin.

- High pressure in upper half of body, low (or normal) pressure behind stenosis
- Pressure overload of the LV … LV hypertrophy
- Interscapullar systolic murmur

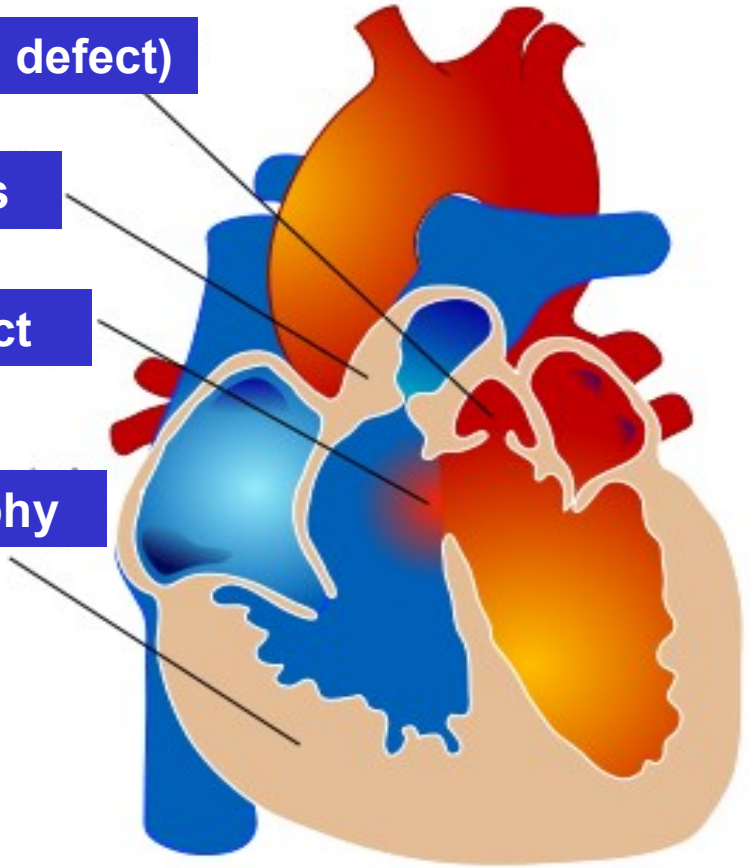


- Diagnostics: trans-oesophageal echo (TEE)
  MRI - angiography
- Complications: cardiac failure,
  infectious endocarditis,
  aneurysm of the brain arteries
- Therapy: surgery

Fallot tetralogy

Combined obstructive and shunt defect:

- Overriding aorta (at septal defect)
- Pulmonary stenosis
- (Subaortic) ventricular septal defect
- Right ventricular hypertrophy



Fallot tetralogy

Right-to-left ventricular shunt

Low oxygenation – cyanosis, dyspnoe – early in newborns,

Polyglobulia

Complications: hypoxemia, brain ictus in children, infectious endocarditis

If not treated – an early death during several years

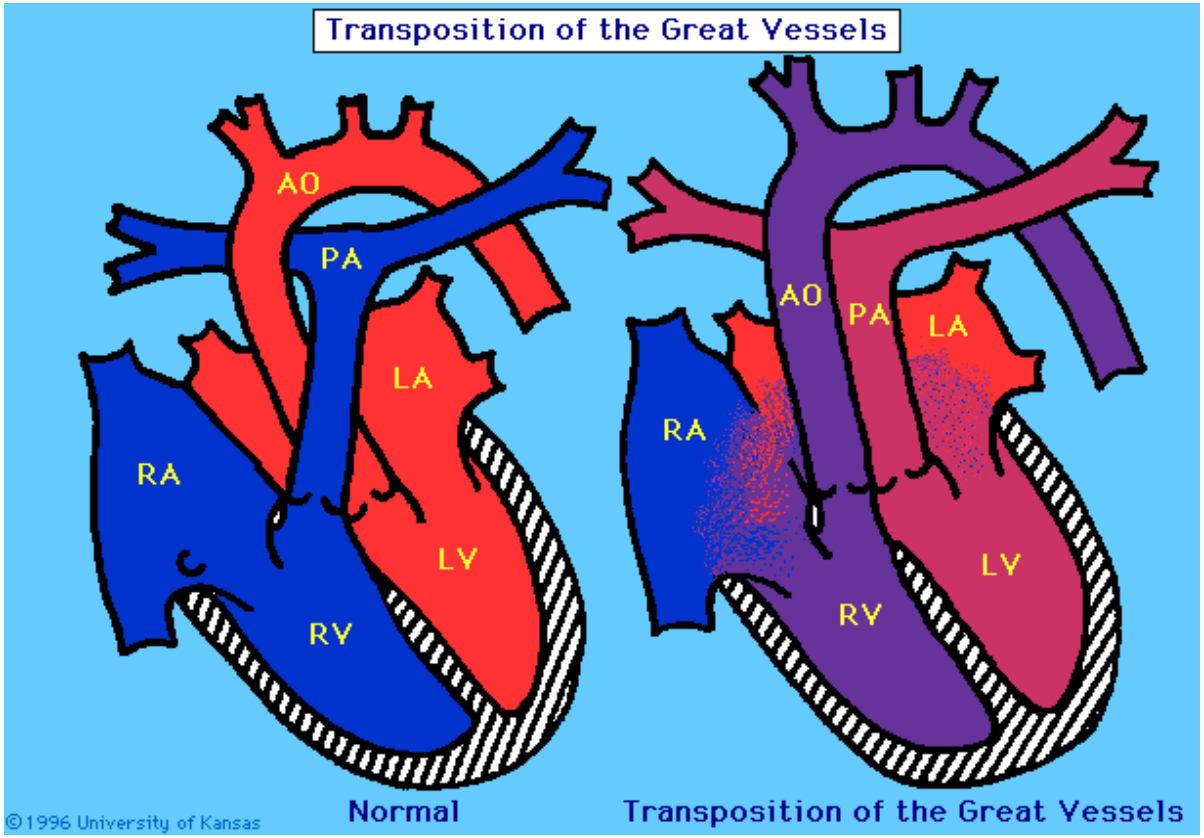
Systolic murmur in 2nd intercostal space on the left side

Dg.: ECHO

Therapy: surgical correction

Transposition of the great vessels

- **Dislocation defect**
- **Aorta leads from RV, pulmonary artery leads from LV**



Transposition of the great vessels

- Dislocation defect
- Aorta leads from RV, pulmonary artery leads from LV
- **Isolated defect is incompatible with a life. Therefore it is usually accompanied by a shunt defect (foramen ovale patens, ventricular septal defect, persistent truncus arteriosus)**
- **Risk factor: Preexisting DM in the mother (as well as for aortic coarctation)**

Transposition of the great vessels

- **Cyanosis, dyspnoe in newborns, cardiac failure, systolic murmur in precordium**
- **Diagnostics: echo, heart catheterization**
- **Poor prognosis – the death in first weeks of a life**
- **Therapy: switch surgery**