HEART DISEASES
Myocarditis, cardiomyopathy

Pavel Maruna
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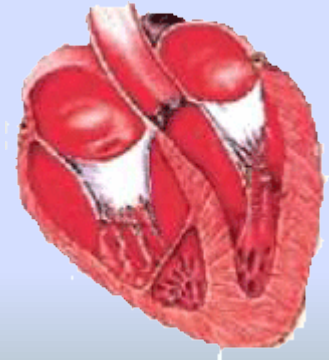
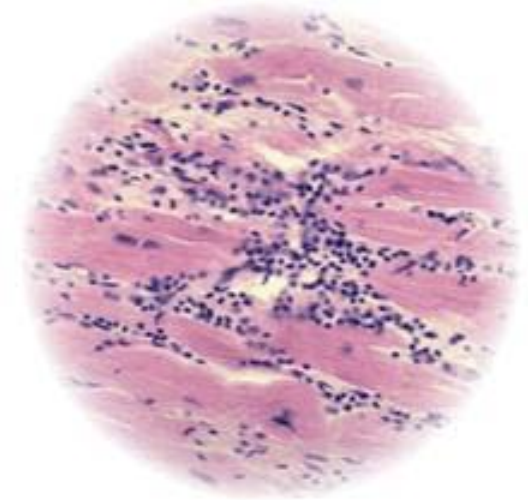


Myocarditis

Etiology: infection + (auto)immunity

- rheumatoid fever
- diphteria
- streptococcal infection
- mycoplasma
- salmonelosis
- Weil dis. (leptospirosis)
- ricketsia
- influensa, polio, parotitis, CMV
- Chagas dis. (trp. crusii)
- systemic dis. of connective tissue
- immunocomplex. vasculititis
- Fiedler idiopatic myocarditis (virosis ?)



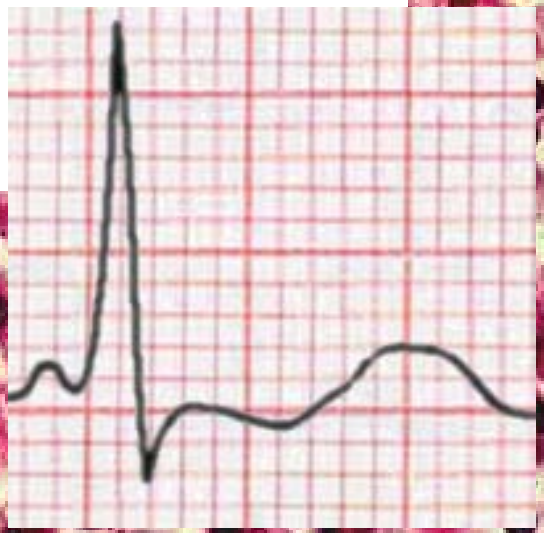
Myocarditis

Manifestation:

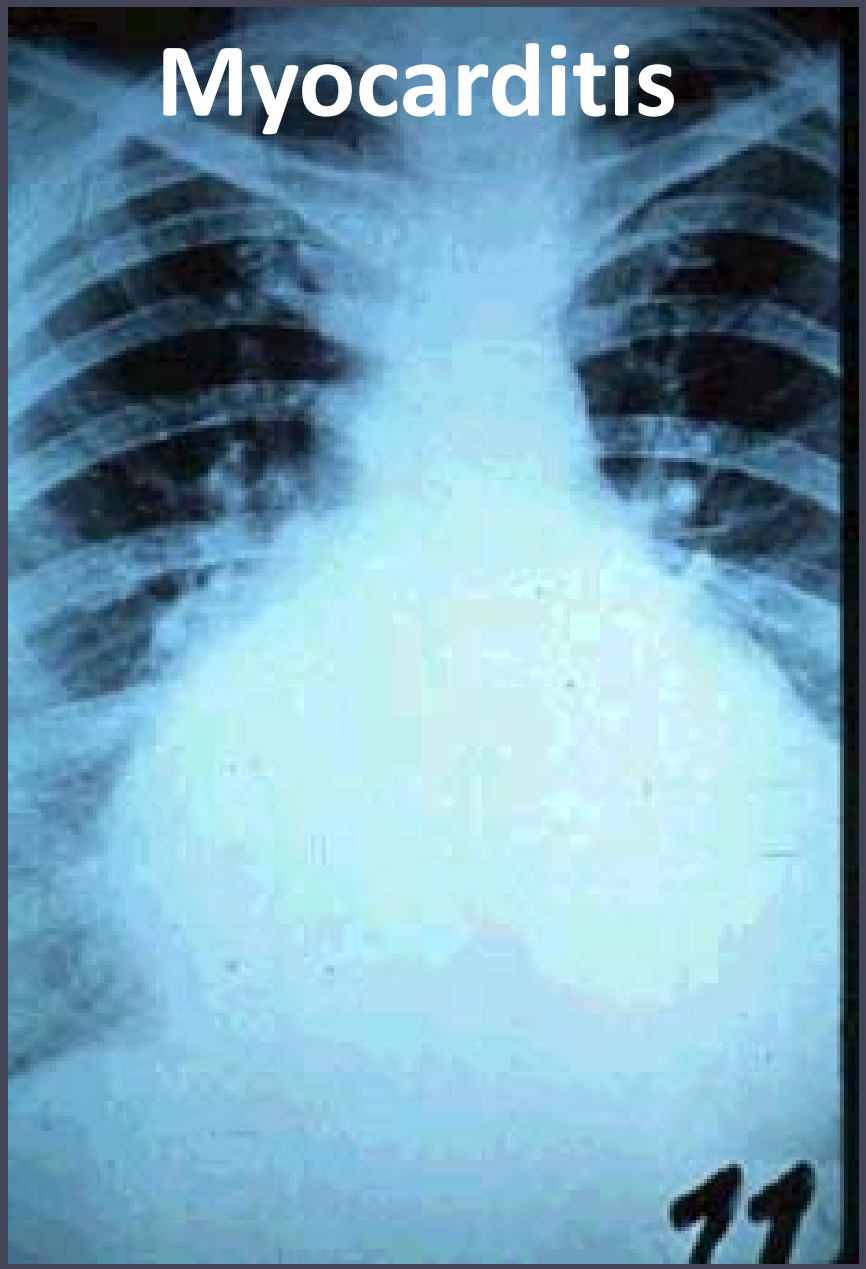
- ♥ asymptotic
- ♥ chest pain, dyspnoe, palpitation
- ♥ arrhythmia
- ♥ cardiac failure
- ♥ ECG: variable finding (PQ int., ST-T, blocks), “myocarditic curve”

Diagnosis:

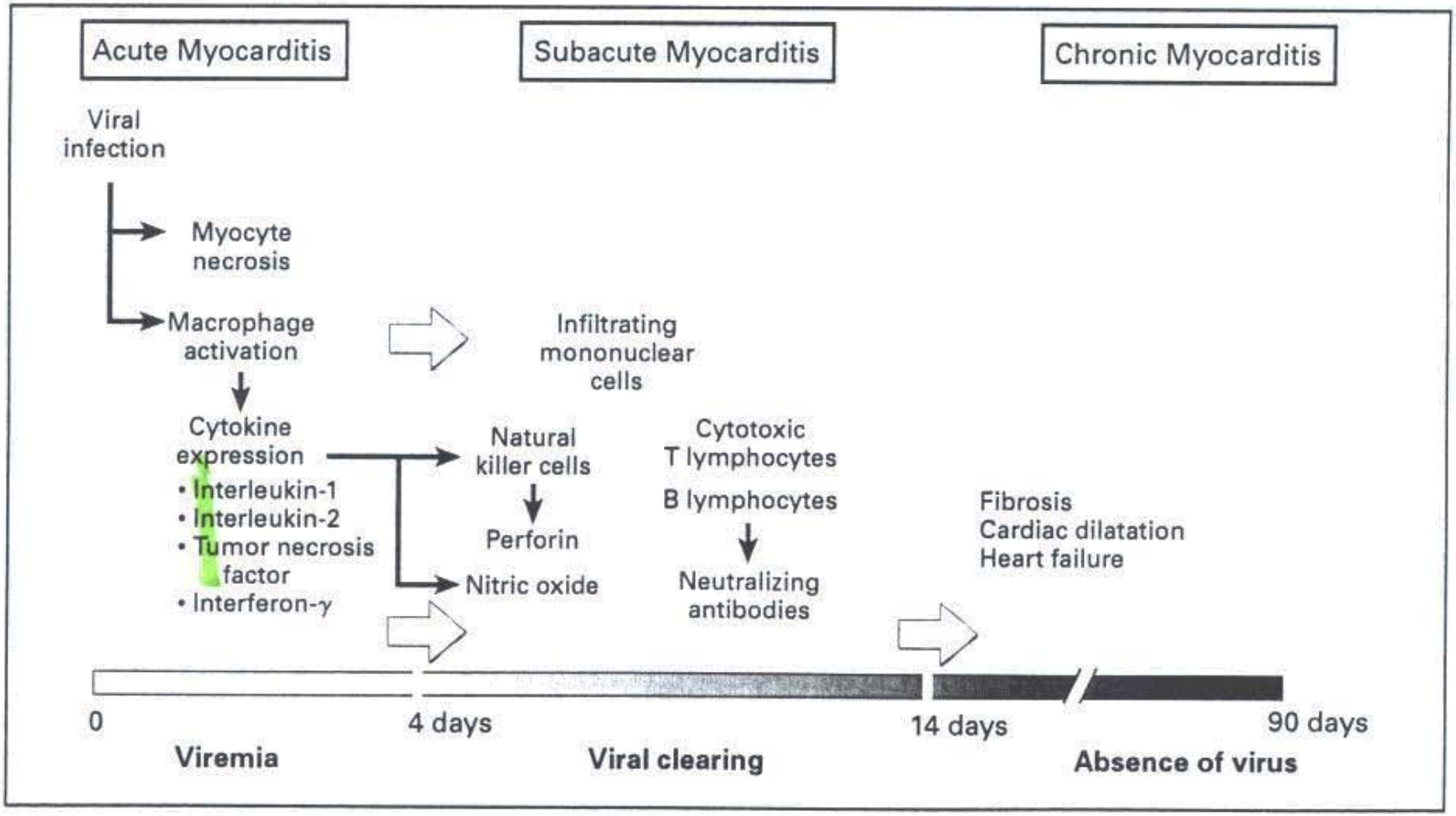
difficult – echocardiography



Myocarditis



Myocarditis

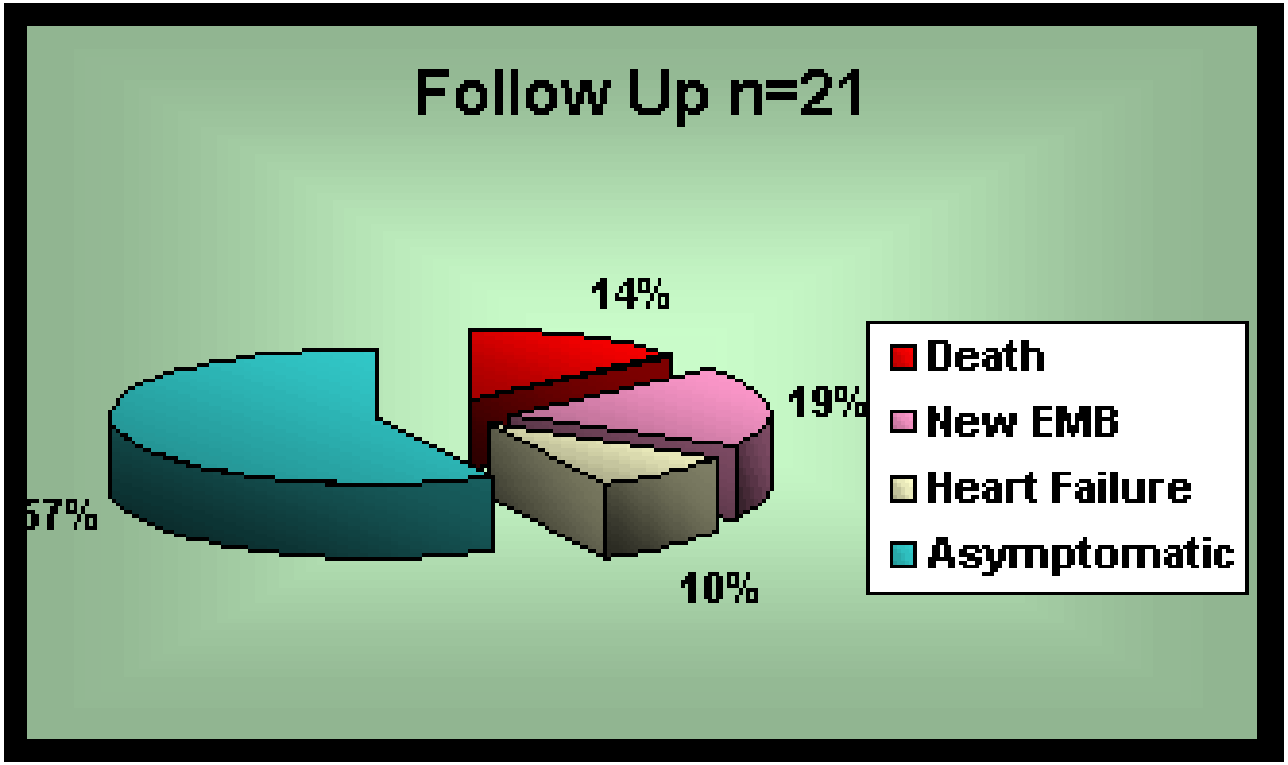


Time Course of Experimental Viral Myocarditis in Mice.
Adapted from Kawai[11] with the permission of the publisher. The timeline is not drawn to scale.

Myocarditis

Prognosis:

recovery / cardiac failure / latent development of dilated CM



Cardiomyopathy

Definition:

= chronic disorder of myocardium with abnormal ventricular both function and morphology

weakening of the heart muscle or a change in heart muscle structure

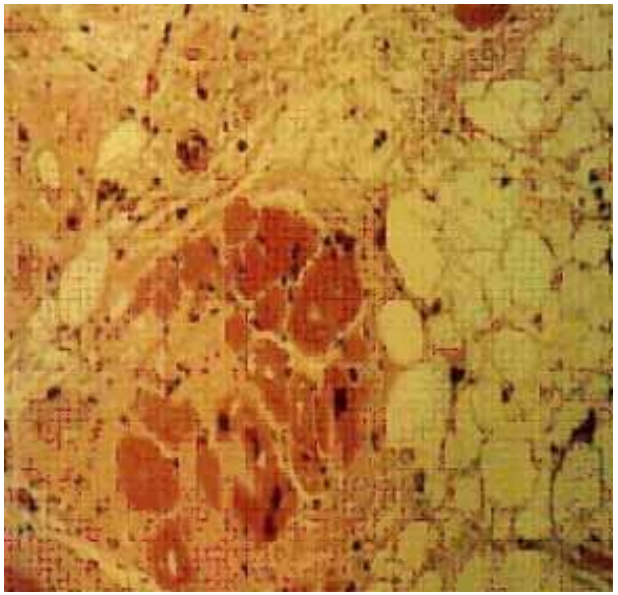
prolonged course, slow progression

Pathogenesis:

"universal" reaction of cardiac muscle on various noxa

→ inflammation, hypertrophy, degeneration, necrosis, fibrosis

→ accumulation of lipids, glycogen, amyloid



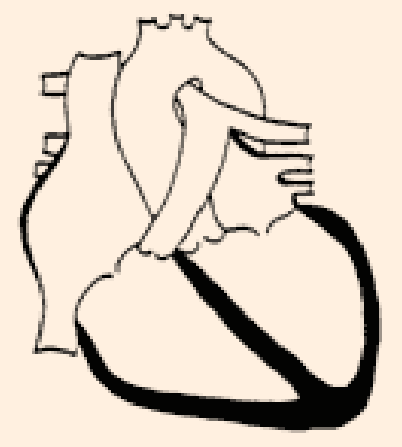
Lipoid deposits in myocardium

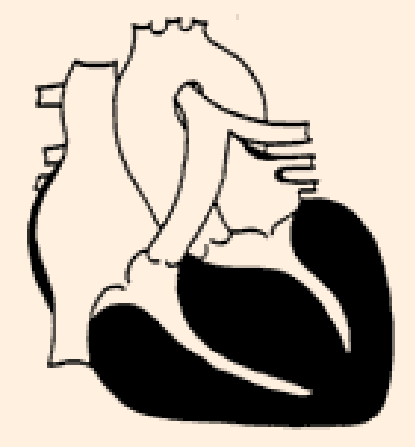
Cardiomyopathy

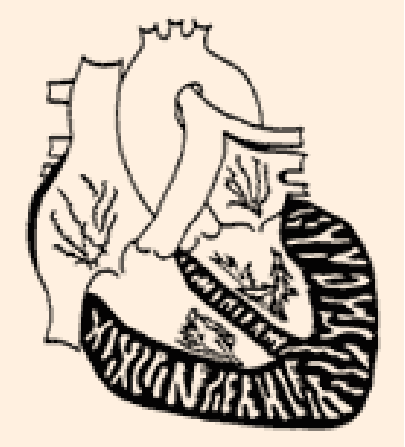
Primary:

Genetic factors, worse prognosis
(must be excluded ischemia, hypertension, congenital + acquired cardiac defects)

Cardiomyopathy



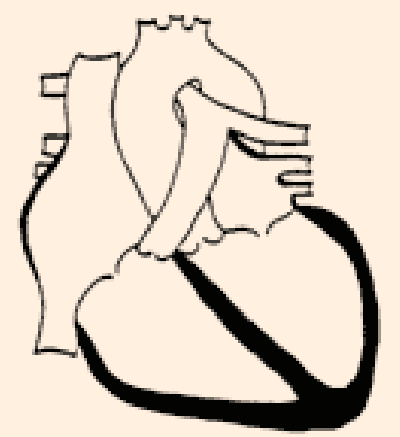


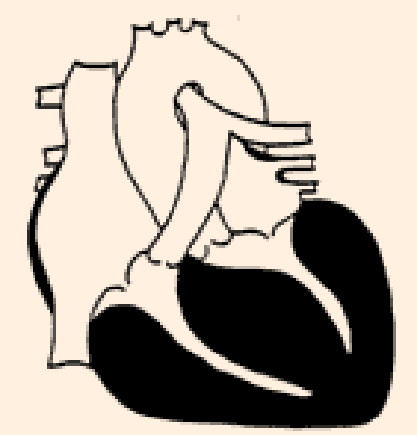


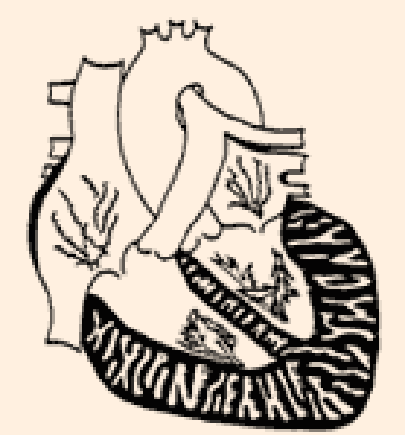
Dilated CM

- destruction of muscle fibers
- dilatation without hypertrophy

Cardiomyopathy



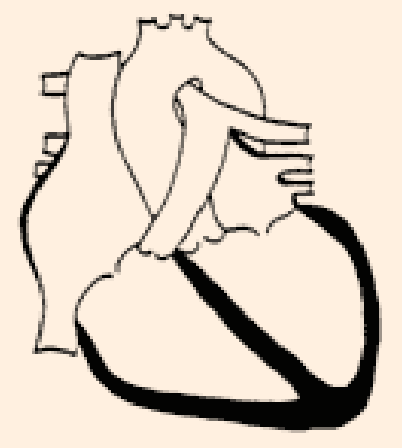


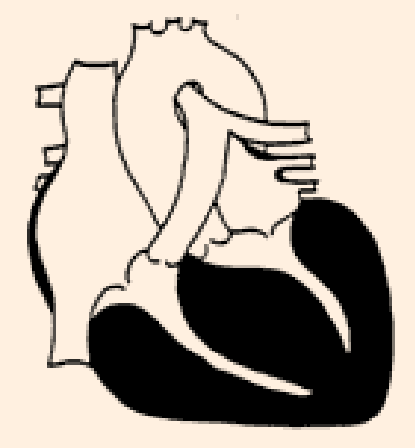


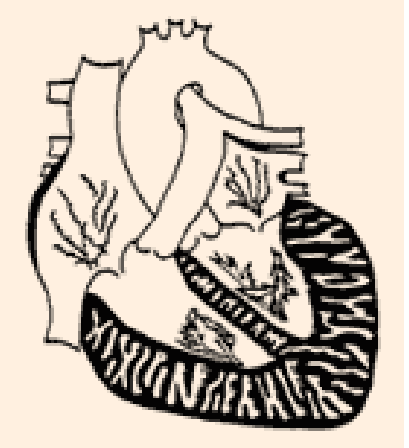
Hypertrophic CM

- asymmetric hypertrophy
- obstruction of LV offtake

Cardiomyopathy







Restrictive CM

- **subendocard. fibrosis**
- **arrhythmia**

TABLE 1

Characteristics and prevalence of the cardiomyopathies vs hypertension, a common cardiovascular condition

DISORDER	PRESENTING SYMPTOMS	ECHOCARDIOGRAPHIC FINDINGS	PREVALENCE	GENE IDENTIFIED
Hypertrophic cardiomyopathy	Chest pain Arrhythmias Dyspnea	LV hypertrophy	1:500	10 genes, > 200 mutations
Dilated cardiomyopathy	Heart failure Arrhythmias	RV or LV dilatation	1:2,500	15 genes, > 20 mutations
Arrhythmogenic RV cardiomyopathy	Arrhythmias Heart failure	RV dilatation and dysfunction	1:1,000 to 1:5,000?	3 genes, > 8 mutations
Restrictive cardiomyopathy	Dyspnea Heart failure	LV stiffness Enlarged atria	Unknown	1 gene, 5 mutations
Systemic hypertension	Hypertension Dyspnea	Can cause LV hypertrophy	1:4	Mostly complex traits

LV = left ventricular, RV = right ventricular

Cardiomyopathy

Secondary:

infectious

- **bacterial**
- **viral (coxsackie)**
- **ricketsia**
- **mycosis**
- **parasitic (Chagas dis.)**

toxic (alcohol, Co, narcotics, psychofarmacs, adriamycin, prokainamid)

endocrine / metabolic (↓T4, ↑T4, ↑GH, uremia, ↓vit.B1, K, Mg)

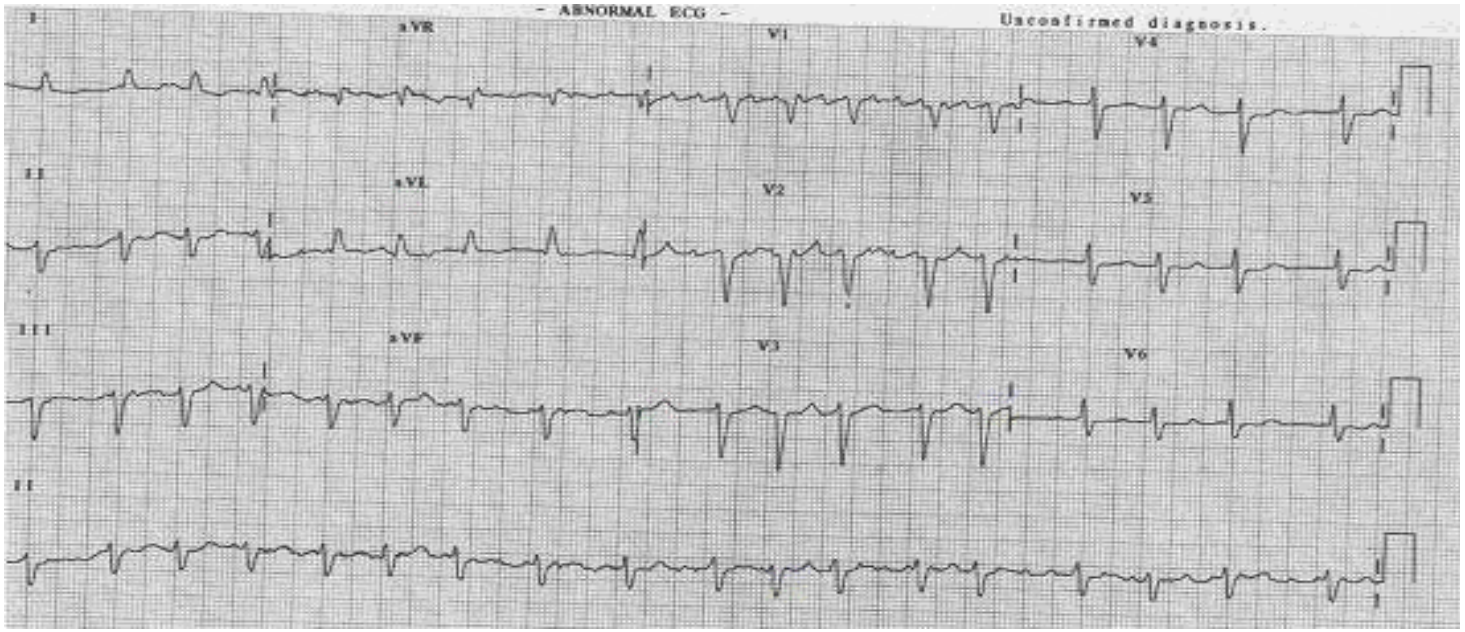
allergy, autoimmunity (immunocomplex., SLE, sarkoidosis…)

Cardiomyopathy

ECG:

SVES, VES, atrial fibrillation
RBBB, LBBB
T wave aplanation / inversion
LV hypertrophy (♂ > 400 g, ♀ > 385 g)

It is unusual for patients with cardiomyopathy to have a normal ECG

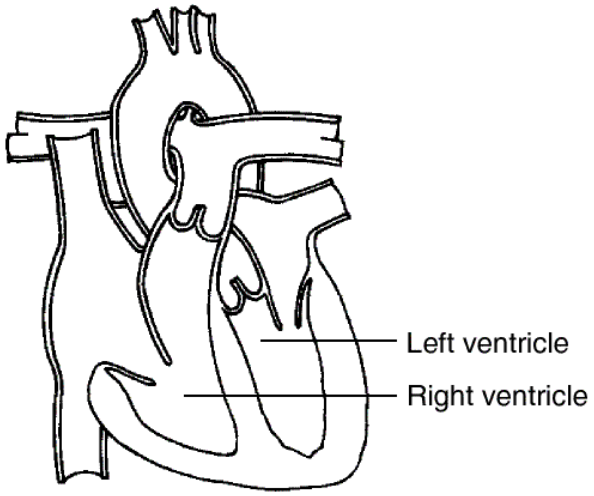


Dilated (congestive) CM

Characteristics:

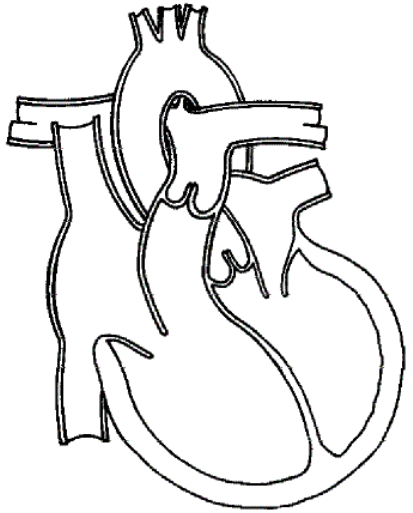
- ♥ heart dilation (without hypertrophy), diffuse hypokinesis (systolic + diastolic dysfunction)
- ♥ (passive) pulmonary hypertension, right heart failure
- ♥ arrhythmia, blocks

Normal Heart



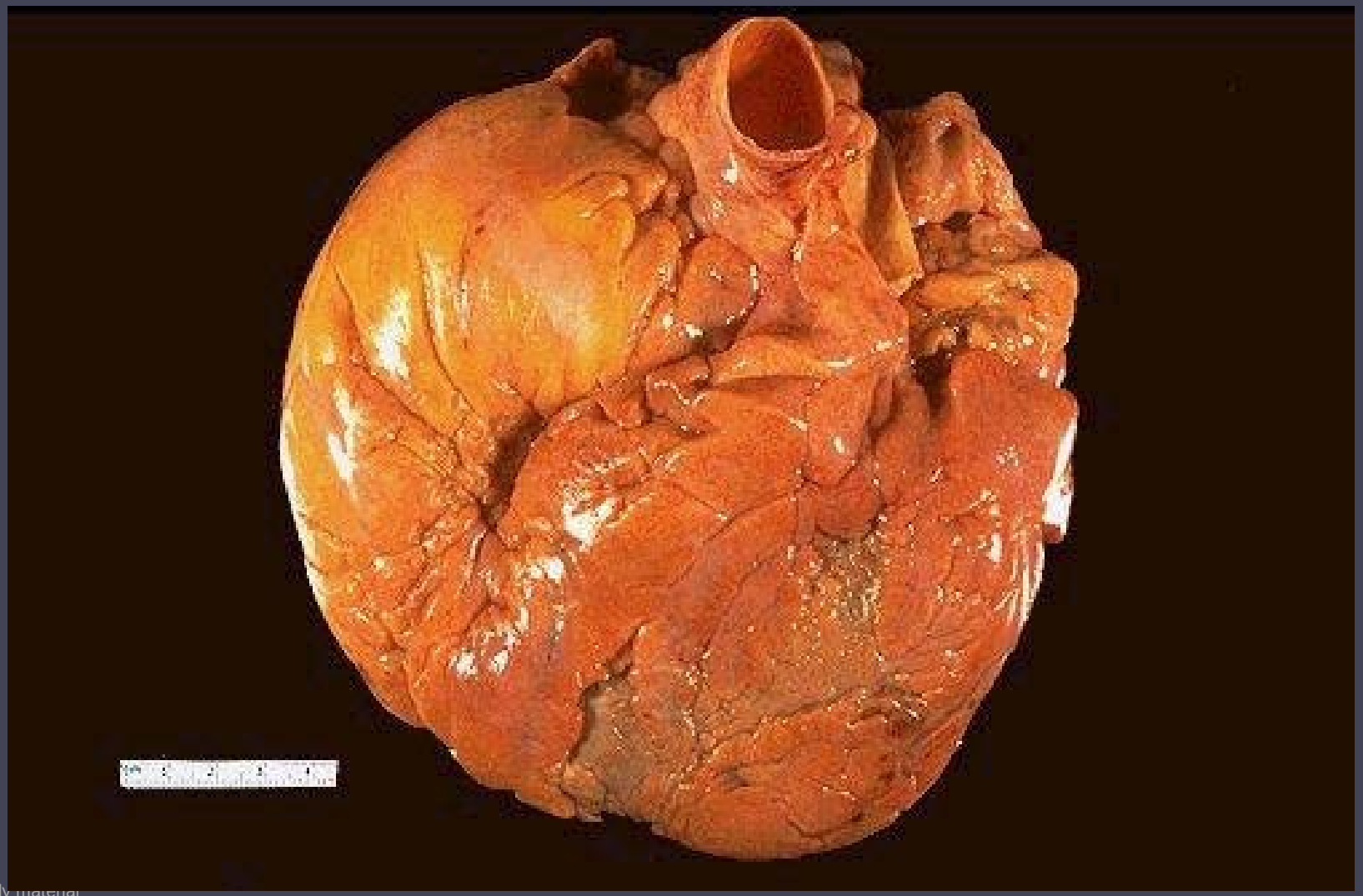
Heart chambers relax and fill, then contract and pump.

Heart with Dilated Cardiomyopathy



Muscle fibers have stretched. Heart chamber enlarges

Dilated (congestive) CM



Dilated CMP

dilatation **of all heart with decreased contractility**
Decreased *systolic* function

low EF
high residual volume in the ventricle
increased EDV and lung congestion
decreased systolic volume and pressure

Dilated CMP

Symptoms typical for heart failure, symptoms both of
low cardiac output*, and *congestion

Frequent *arrhytmias* and
***thromboembolic* complications**

Relative valvular *regurgitation*

Causes of dilated CMP

*genetic (cytoskeletal proteins, myopathies…) – 30-50 %

- intoxication (alcohol, cytostatics, cobalt, drugs…)
- metabolic diseases
- autoimmune postinfectious mechanisms (sequelae of viral myocarditis, e.g. Coxsackie)
- other…

Dilated (congestive) CM

Links:

- **alcoholism (+ malnutrition, ↓vitamin., hepatopathy…)**
- **coxsackie B (e.g. intrauterine infection)**
- **hereditary factors (...to examine relatives)**
- **drug factors (ATB, sympathomimetics)**

Alcoholic Cardiomyopathy* : Incidence, Clinical Characteristics, and Pathophysiology

Mariann R. Piano

Chest 2002;121;1638-1650

ALCOHOL

Alcohol consumption ⇩ *> 90gms > 5 years*

- Apoptosis (either directly via alcohol or indirectly via ↑ NE levels)
- ↓ synthesis and/or accelerated degradation of contractile proteins
- ↓ myofilament Ca^{2+} sensitivity
- Intrinsic myocyte dysfunction due to mitochondrial and sarcoplasmic dysfunction (due to Ca^{2+} overload, fatty ethyl esters or NE)

⇩

Cell drop out and weakly contracting myocytes

⇩

Decreased cardiac output

⇩

- LV dilation to increase EDV (preload) to compensate for ↓ cardiac output, however this is may be accompanied by wall thinning due to cell drop out
- Hypertrophy of normal myocytes to compensate for weakly contracting neighboring myocytes

Continued drinking ⇩ *> 15 years*

- Progressive LV dilation and wall thinning
- Activation of other neurohormonal systems
- Signs and symptoms of heart failure

FIGURE 2. Proposed hypothetical schema for the pathogenesis of ACM. gms = grams; NE = norepinephrine.

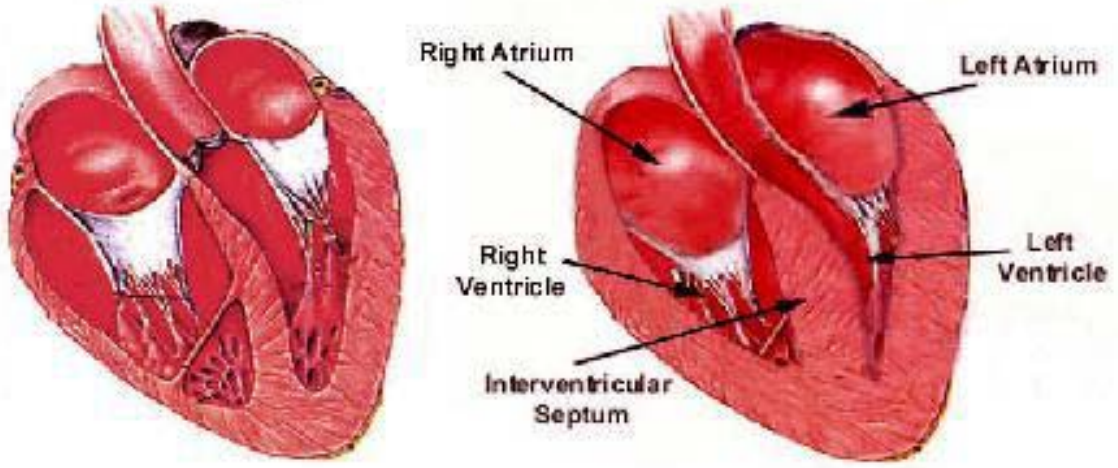
Hypertrophic (obstructive) CM

= subaortic stenosis = subvalvular idiopatic aortic stenosis

Characteristics:

The influence of catecholamines on fetal heart or ↑ catecholamine receptors in fetus

Often AD heredity (to examine relatives)



Hypertrophic (obstructive) CM

♥ asymmetric hypertrophy LV > septum > RV (with ECG picture)
♥ microscopy: disorganization of musculature, islets of fibrosis
♥ vault of hypertr. septum >>> obstruction of aortic intake

→ normal systolic function, low diastolic compliance
→ ventricular arrhythmia (risk of sudden death)
→ dizziness, syncope
→ intolerance of strain, dyspnoe
→ palpitation

SUDDEN DEATH

Hypertrophic CMP

abnormal *hypertrophy* of the myocardium, mainly of LV – subaortic stenosis) w/o stimulus

Normal *systolic* function
Impaired ejection of the blood due to the obturation

Disturbed *ventricle filling,* diastolic dysfunction (increase in EDV)

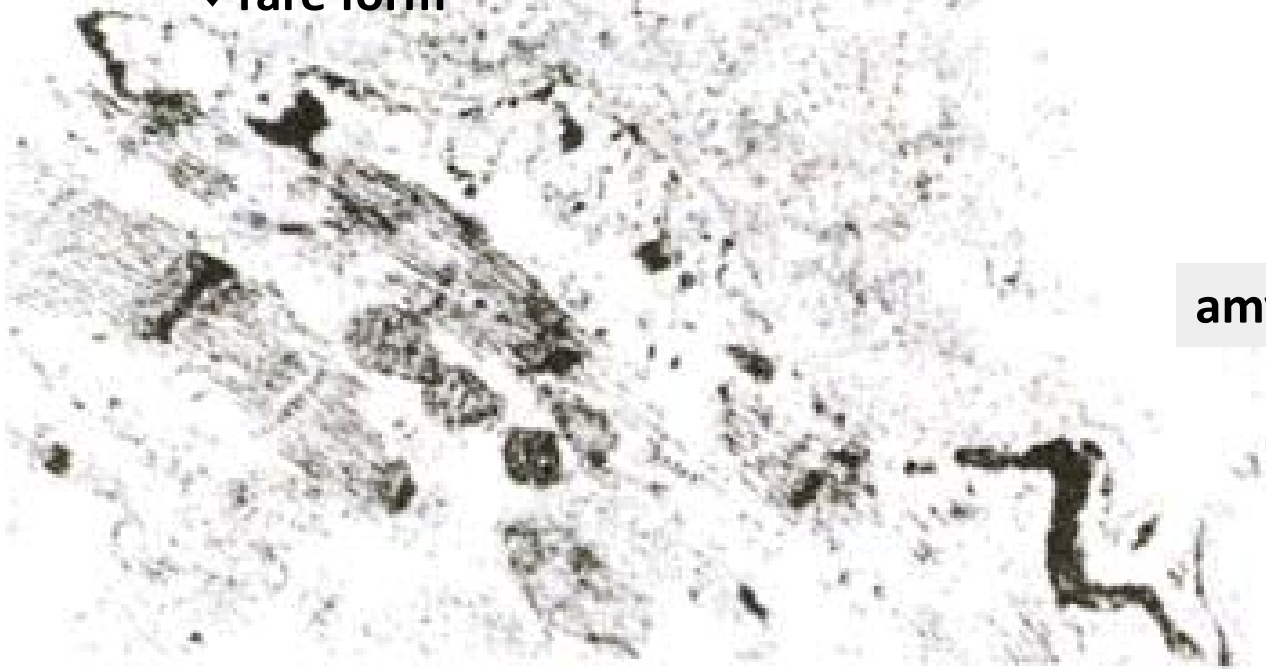
Causes

- various mutation of several genes in the sarcomera – actin, myosin, tropomyosin…
- up to 90 % AD hereditance – test the family!
- incidence up to 1:500

Restrictive CM

Characteristics:

- ♥ subendocardial fibrosis (event. eosinophil infiltration)
- ♥ frequent arrhythmia
- ♥ heart is normal in size or only slightly enlarged
- ♥ rare form



amyloid deposits

Restrictive CM

Symptoms:

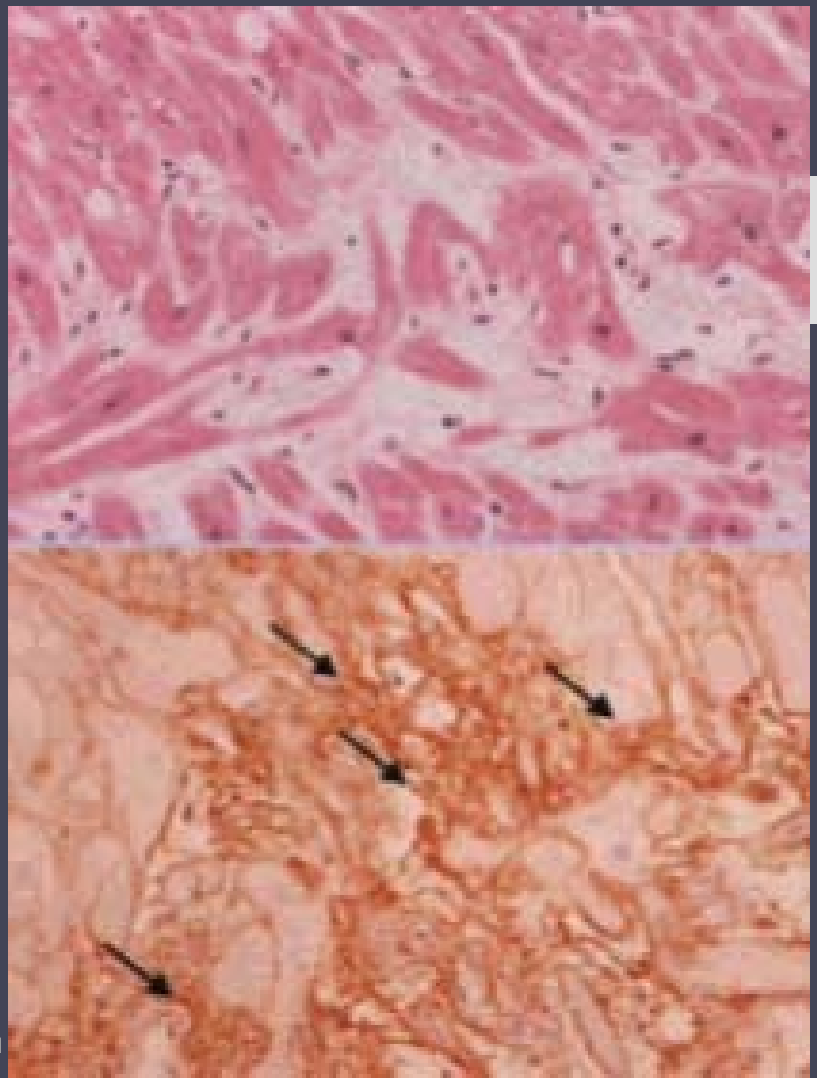
- ♥ excessive tiredness (fatigue), poor tolerance of exercise
- ♥ cough - difficulty breathing
- ♥ palpitation, syncope - arrhythmia

Restrictive CM

Prognosis:

People with restrictive CM may be candidates for heart transplant.
Prognosis is dependent on the underlying cause but it is usually poor.
Average (mean) survival after diagnosis is 9 years.

Restrictive CM

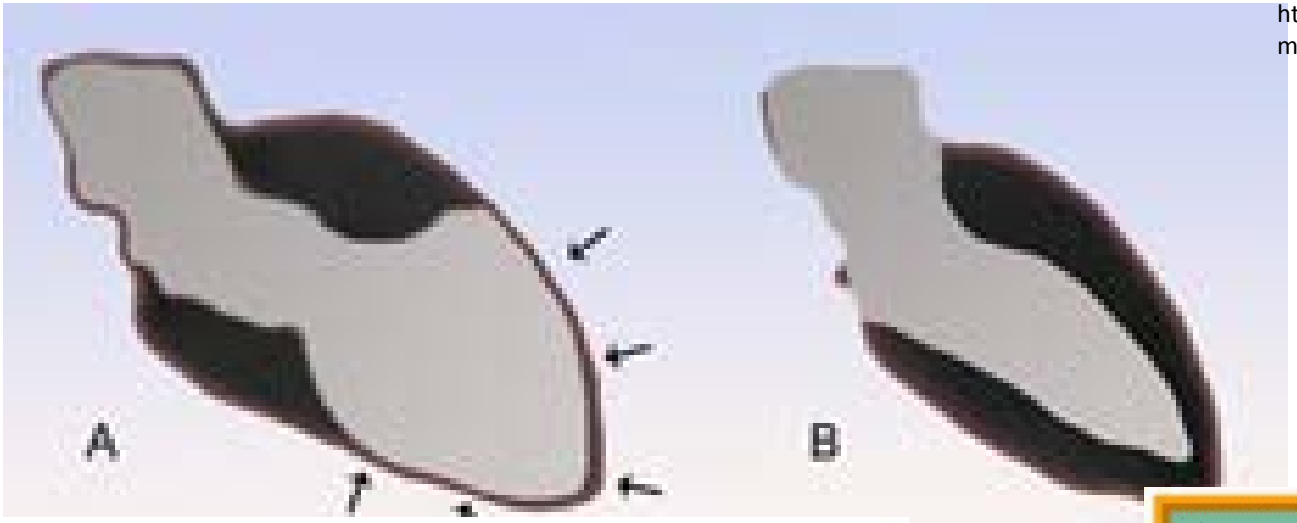


eosinophil fibrillar structures in myocardial interstitium

Stress CMP – broken heart syndrome, tako tsubo

- nonischemic CMP with sudden narrowing of myocardium usually around the apex with contractility disturbance
- **during the contraction the apex area remains akinetic (balloon-like) while the base area contracts normally (narrowing)**
- reminds the octopus trap – tako tsubo
- described first in 1991
- rare, estimated 12 000 in USA/year

Heart changes in stress CMP



http://en.wikipedia.org/wiki/Takotsubo_cardiomyopathy

tako = chobotnice
tsubo = nádoba

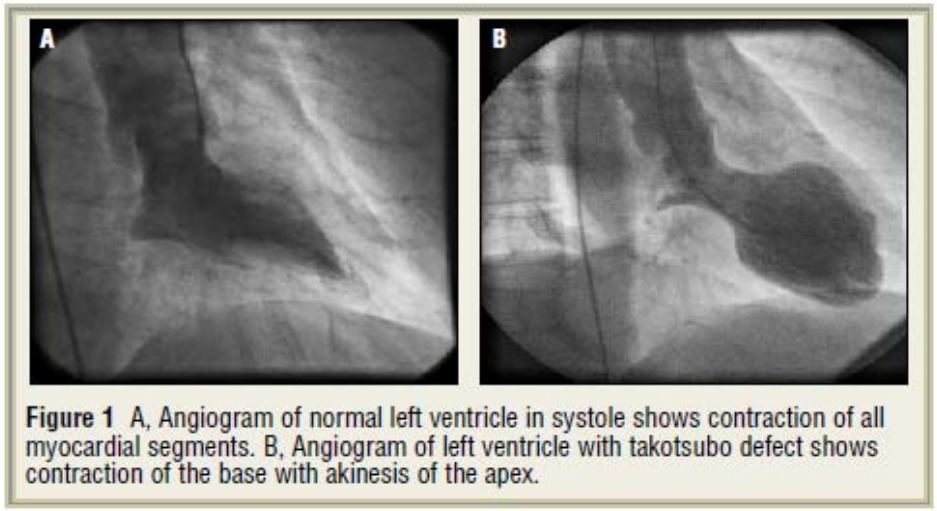


Figure 1 A, Angiogram of normal left ventricle in systole shows contraction of all myocardial segments. B, Angiogram of left ventricle with takotsubo defect shows contraction of the base with akinesis of the apex.

The "Broken Heart Syndrome": Understanding Takotsubo Cardiomyopathy

Clinical picture of stress CMP

- occurs **mainly in postmenopausal women** following emotional or physical stress – „broken heart“
- acute, severe symptoms – dyspnea, lung edema, ecg changes, arrhytmia, chest pain… - **resembles acute heart attack (infarction)**
- no signs of coronary arteries narrowing and/or CHD risk factors
- risk of heart rupture
- **usually has tendency for spontaneuous improvement** in days or weeks

Table 2 Emotional and physical stressors associated with takotsubo cardiomyopathy

Emotional stressors
Unexpected death of relative or friend[1,7]
Domestic abuse[1]
Confrontational argument[1,7]
Catastrophic medical diagnosis[1]
Devastating business[1]
Armed robbery[7]
Gambling losses[1]
Surprise party[7]
Surprise reunion[7]
Car accident[7]
Fear of procedure[7]
Fear of choking[7]
Court appearance[7]
Public performance[7]
Physical stressors
Exacerbated systemic disorders[1]
Noncardiac invasive procedures[1,13]
Exhausting physical effort[1,5]
Asthma attack[1]
Pneumothorax[5]
Ventricular fibrillation[5]
Cold exposure[5]

Theories of the pathogenesis

- too much catecholamines
- temporary multiple vasospasms
- why mainly women ?
  why mainly the apex ?