

Myocarditis and Cardiomyopathies

L.V. Bogun, N.I. Yabluchansky, F.M. Abdueva, O.Y. Bichkova, A.N. Fomich, P.A. Garkavyi, A.L. Kulik, N.V. Lysenko, N.V. Makienko, L.A. Martimyanova, I.V. Soldatenko, E.E. Tomina

Department of Internal Medicine

Faculty of Medicine

Kharkiv V.N. Karazina National University

Lecture for 5 course, update 2013

Myocarditis

Definition

- Myocarditis is an inflammatory disease of the heart muscle, diagnosed by established histological, immunological, and immunohistochemical criteria (the World Health Organization/International Society and Federation of Cardiology, 1995)

Epidemiology

- The true incidence of myocarditis is unknown because the majority of cases are asymptomatic
- Involvement of the myocardium has been reported in 1 to 5 % of patients with acute viral infections
- Autopsy studies have revealed varying estimates of the incidence of myocarditis.
- A 5% prevalence of active myocarditis was reported in a high-risk group of 186 sudden, unexpected medical deaths in children

RISK FACTORS

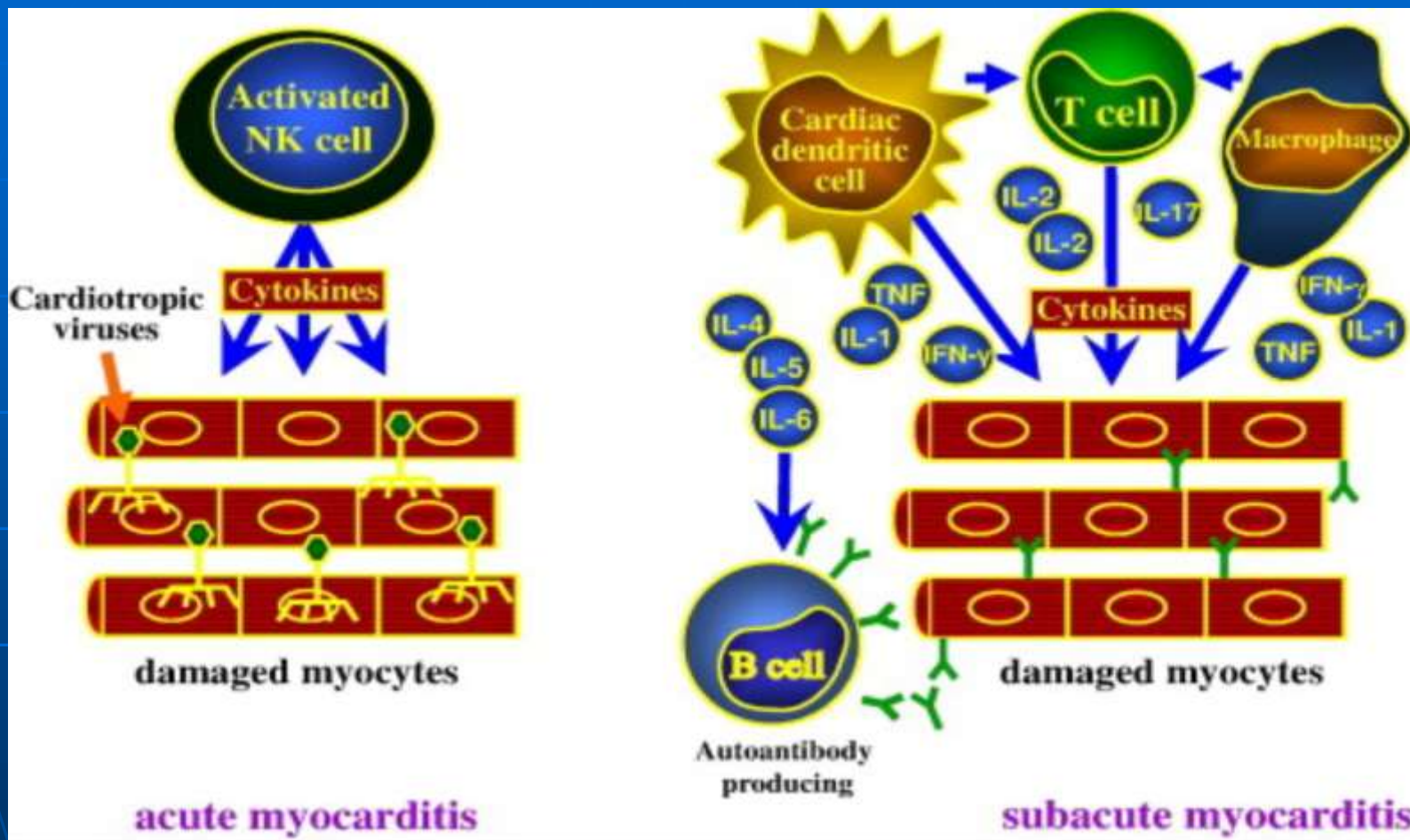
- Certain groups appear to be at increased risk of virus-induced myocarditis, and the course may be hyperacute
 - Young males
 - pregnant women
 - children (particularly neonates)
 - immunocompromised patients (HIV)

Etiology of Myocarditis

■ Infection

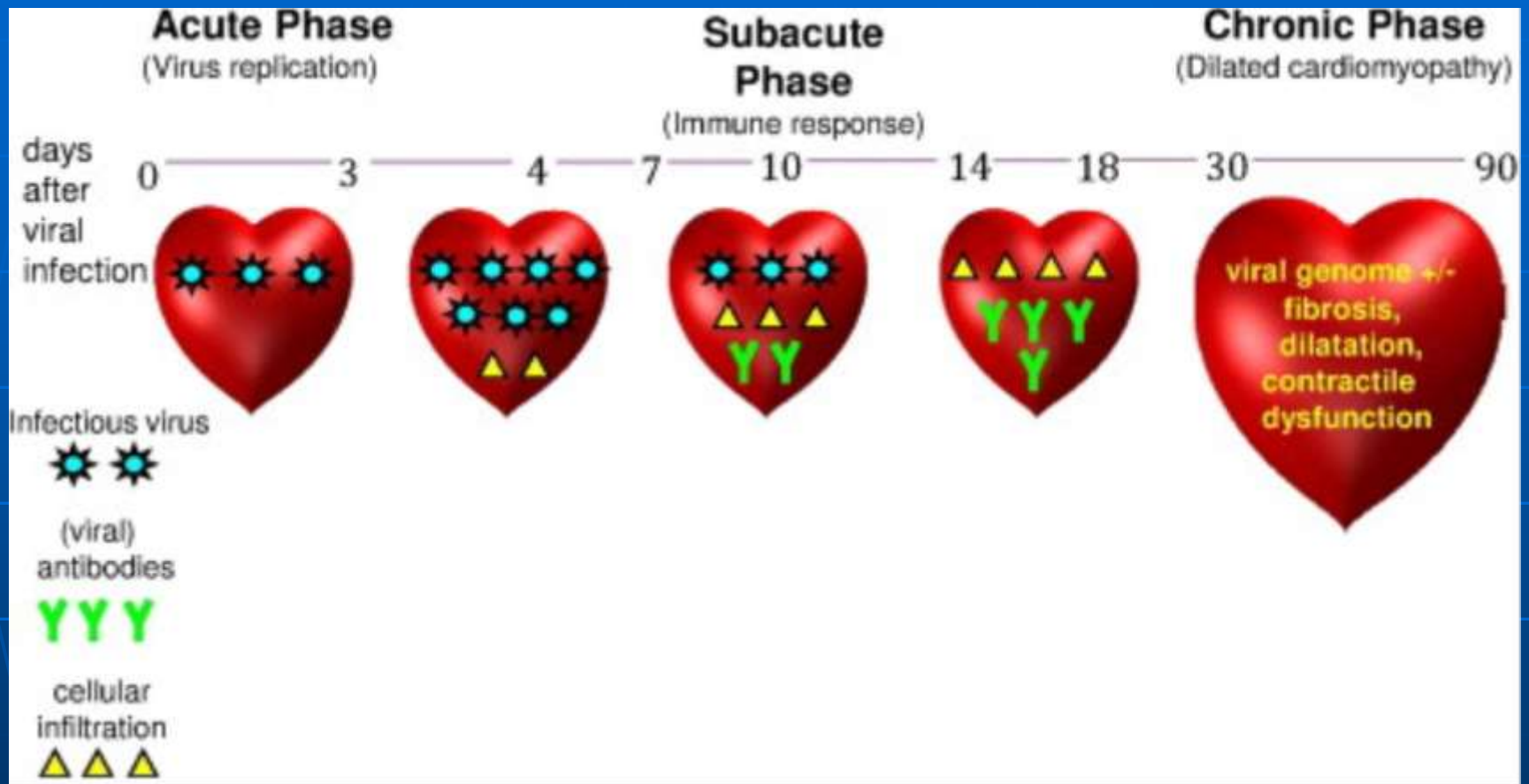
- **Viral** (most frequently identified viruses were enteroviruses (including coxsackievirus) until the 1990s, but now parvovirus B19 and Human Herpes Virus 6 are more common)
- **Bacterial, rickettsial, spirochetal**
- **Protozoal**
- **Fungal**
- **Autoimmune disorders** (celiac disease, systemic lupus erythematosus, Wegeners granulomatosis, giant cell arteritis, Kawasaki's, and Takayasu arteritis)
- **Drugs** (antibiotics, sulfonamides, smallpox vaccine, tetanus toxoid, tricyclic antidepressants)
- **Toxins** (Cocaine, phenytoin, ethanol)
- **Parasites** Chagas, Toxoplasmosis, Trichinosis

Pathophysiology of Viral Myocarditis



Pathophysiology of viral myocarditis: after viral entry, virus replication leads to acute injury of the myocytes (acute myocarditis) and to activation of the host's immune system (subacute myocarditis). IFN = interferon; IL = interleukin; TNF = tumor necrosis factor

Time Course of Viral Myocarditis



Time course of viral myocarditis in 3 phases (derived from murine models). The acute phase of myocarditis takes only a few days, whereas the subacute and chronic phase covers a few weeks to several months.

Clinical presentation

Myocarditis is preceded by

- flu-like symptoms (chills, fever, headache, muscle aches, general malaise)
- gastrointestinal symptoms (decreased appetite, nausea, vomiting, and diarrhea)

Cardiac manifestations of myocarditis appear a few hours to a few days after the initial signs and symptoms.

Cardiac symptoms consist of

- symptoms of heart failure,
- chest pain due to pericardial irritation
- symptoms associated with heart block and arrhythmia.

The possibility of myocarditis must be considered if a patient with **cardiac** symptoms is **febrile**.

Clinical features

- A variety of cardiac symptoms can be induced by myocarditis
- Chest pain may occur, usually due to concomitant pericarditis
- Excessive fatigue or decreased exercise ability may be the initial sign of myocardial dysfunction
- Since **both** ventricles are generally involved, patients **develop biventricular failure**
- Patients present with signs of **right** ventricular failure such as increased jvp, hepatomegaly, and peripheral edema
- If there is predominant **left** ventricular involvement, the patient may present with the symptoms of pulmonary congestion dyspnea, orthopnea, rales, and, in severe cases, acute pulmonary edema

Physical examination

- In addition to the signs of **fluid overload**, the physical examination often reveals direct evidence of **cardiac dysfunction** in symptomatic patients
- S3 and S4 gallops are important signs of impaired ventricular function
- If the right or left ventricular dilatation is severe, auscultation may reveal murmurs of **functional mitral** or **tricuspid insufficiency**
- A pericardial friction **rub** and **effusion** may become evident in patients with **myopericarditis**

Blood studies

- Erythrocyte sedimentation rate elevation - 60%
- White cell count elevation - 25%

Blood Biochemistry

Transient elevation of:

- C-reactive protein (CRP)
- aspartate aminotransferase (AST)
- lactate dehydrogenase (LDH)
- the MB form creatine kinase (CK-MB)
- cardiac troponin T

Viral titers

- Elevation of viral titer in a sample collected in the **acute phase** to at least **four times** that in a sample obtained in **remission** is useful for identify viral infection as the cause.
- Acute and convalescent antibody titers may indicate an active or recent viral infection
- They do not necessarily indicate the etiology of the cardiac abnormalities (most viruses involved in the pathogenesis of myocarditis are **highly prevalent in the population** - 70% of the population in Germany have been tested seropositive for PVB19 Jg G antibodies)

Polymerase chain reaction

- used to demonstrate the presence of viral infection and to detect the viral genome.

Separation of virus in pericardial effusion or cardiac muscle tissue provides direct evidence of myocarditis

Chest X-Ray

- ranges from normal to
- cardiac enlargement - the cardiac silhouette may also be globular when a pericardial effusion is present
- pulmonary congestion

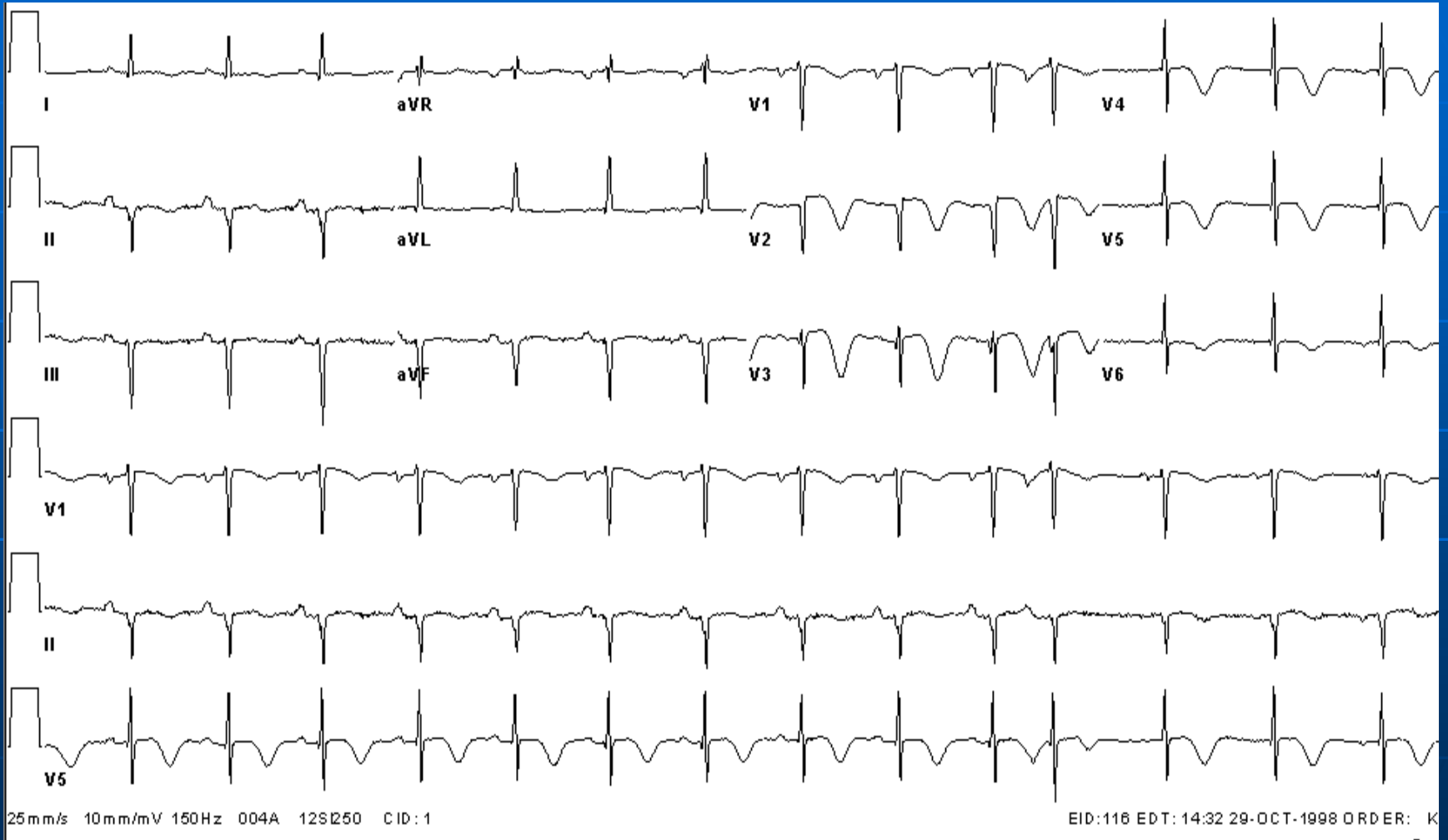
Electrocardiography (I)

- a sensitive and convenient means of diagnosis of myocarditis
- must be timely repeated, since minor abnormalities in the ECG detected initially may become clearer over time
- continuous ECG monitoring is crucial to detect potentially fatal arrhythmias

Electrocardiography (II)

- May be normal or abnormal
- However, the abnormalities are **nonspecific** unless there is pericardial involvement (**myocardial infarction pattern**)
- The changes that may be seen include
 - **sinus tachycardia is most common**
 - nonspecific ST –T abnormalities
 - **supraventricular and ventricular arrhythmias**
 - conduction blocks
 - a gradual increase in the width of the QRS complex is a sign of exacerbation of myocarditis.

Myocarditis



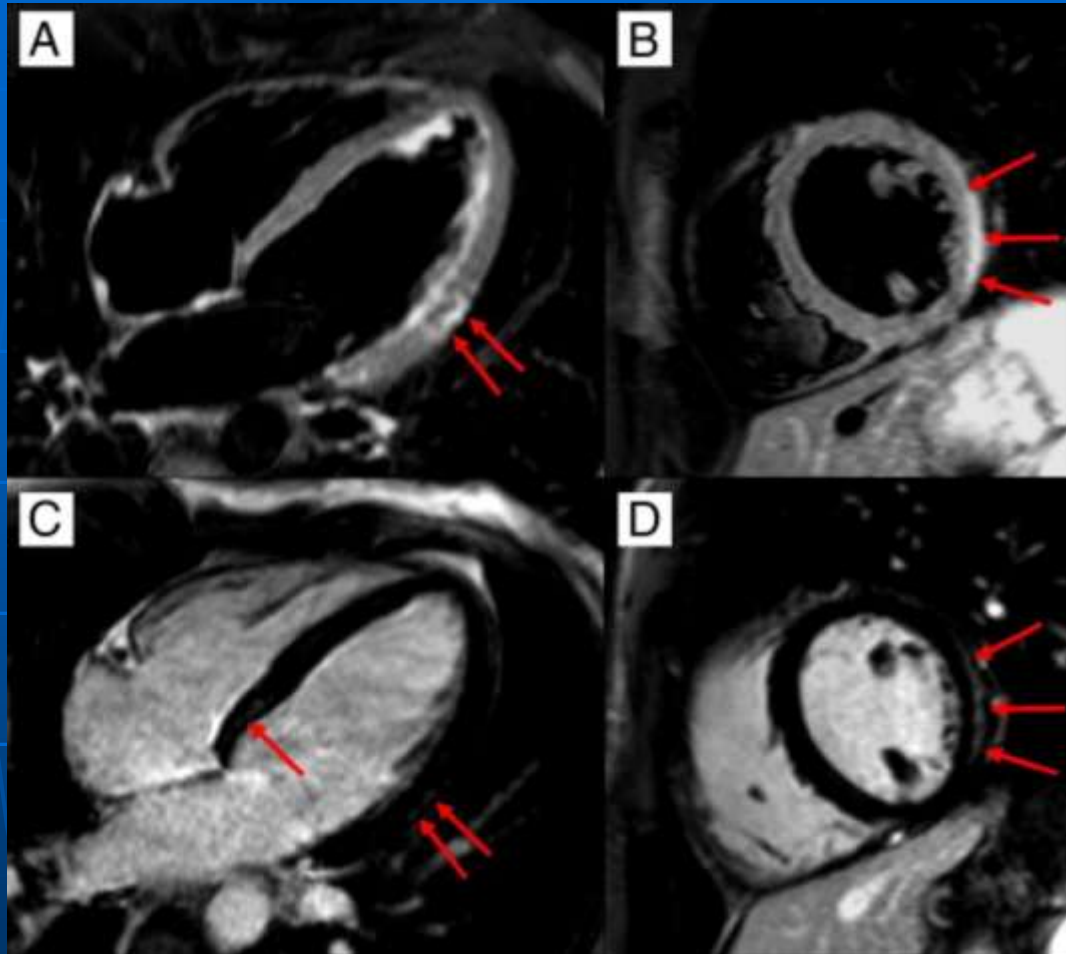
Echocardiography

- Useful tool in managing patients with acute myocarditis
 - **LV systolic dysfunction** is common with segmental wall motion abnormalities/global dysfunction
 - **LV size** is typically normal or mildly dilated
 - **wall thickness** may be increased
 - pericardial **effusion**
 - ventricular **thrombi** may be detected

Magnetic resonance imaging

- Contrast-enhanced MRI, using gadopentate dimeglumine which **accumulates in inflammatory lesions**, can detect the degree and extent of inflammation
- The extent of relative myocardial enhancement correlates with clinical status and left ventricular function

MRI Findings in Patients With Myocarditis



(A) Long-axis and (B) short-axis T2-weighted edema images demonstrating **focal myocardial edema** in the **subepicardium of the left midventricular lateral wall** (red arrows). Corresponding (C) long-axis and (D) short-axis T1-weighted late gadolinium enhancement images demonstrate presence of **typical late gadolinium enhancement** in the **subepicardium of the left midventricular lateral wall and the basal septum** (red arrows).

Endomyocardial biopsy (EMB)

- The gold standard in diagnosis of myocarditis - the definitive diagnosis of myocarditis can be made only by EMB
- A negative EMB does **not rule out** focal myocarditis because of sampling error. This problem is minimized by multiple biopsies

Endomyocardial Biopsy (EMB)

- **RV bioptome permit repetitive sampling**
- **biopsy should be applied early after onset of symptoms to maximize yield - resolution may be seen in four days on serial biopsies**

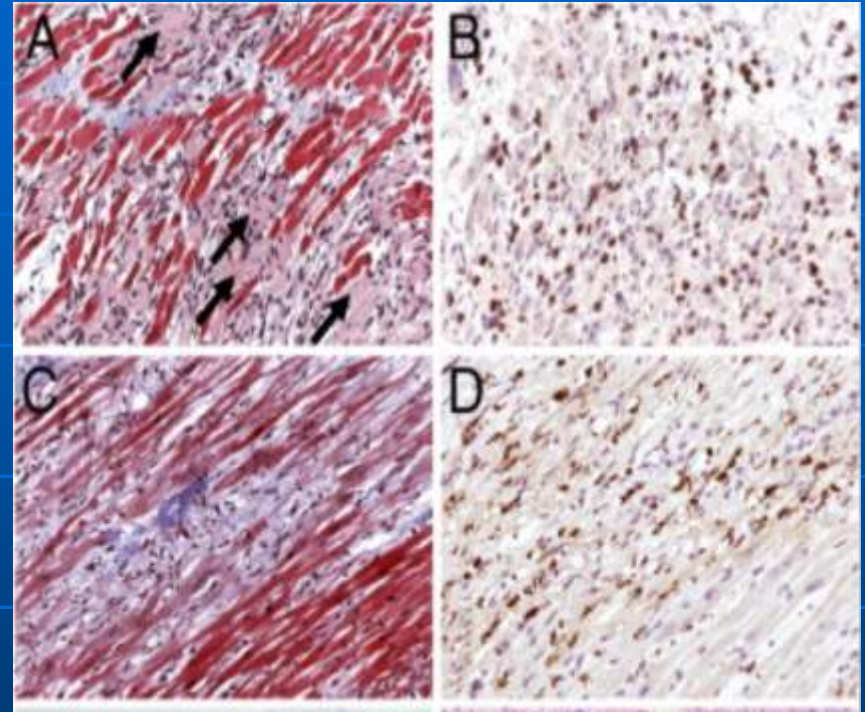
EMB: DALLAS CRITERIA DX OF MYOCARDITIS

- Introduced in 1986 for dx of pericarditis
- Based on endomyocardial biopsy specimens
- **Active** Myocarditis if light microscopy revealed infiltrating **lymphocytes** and **myocytolysis**
- **Borderline or on going** myocarditis if **lymphocyte** infiltration and **NO myocytolysis**
- **Negative for Myocarditis** if **no** lymphocytic infiltrate and **no** myocytolysis

Histopathological Findings in Hearts of Patients With Myocarditis

(A, B) acute myocarditis - numerous **necrotic myocytes** (A, arrows) are associated with **mononuclear cell infiltrates**

(C, D) chronic myocarditis - **inflammatory cells** are mainly present in areas with **fibrosis** (C, blue staining).



Clinical criteria of myocarditis proposed by New York Heart Association (1964; 1973 years)

Relation to the etiological factor : previous infection, proven by clinical and laboratory data (pathogen isolation , immunological tests); allergy, exposure to chemical or physical factors

+

Signs of myocardial lesion:

Major criteria

Minor criteria

Diagnosis of myocarditis is considered in the presence of etiological factors AND the presence of 2 major OR 1 major and 2 minor criteria

Signs of myocardial lesion:

Major criteria

- ECG changes
- Elevated levels of cardioselective enzymes and proteins (CPK, CPK-MB, troponin T)
- Heart enlargement (X-ray, ultrasound)
- Heart failure
- Cardiogenic shock

Minor criteria

- Tachycardia (sometimes bradycardia)
- Weakening of 1st tone while auscultation
- Gallop

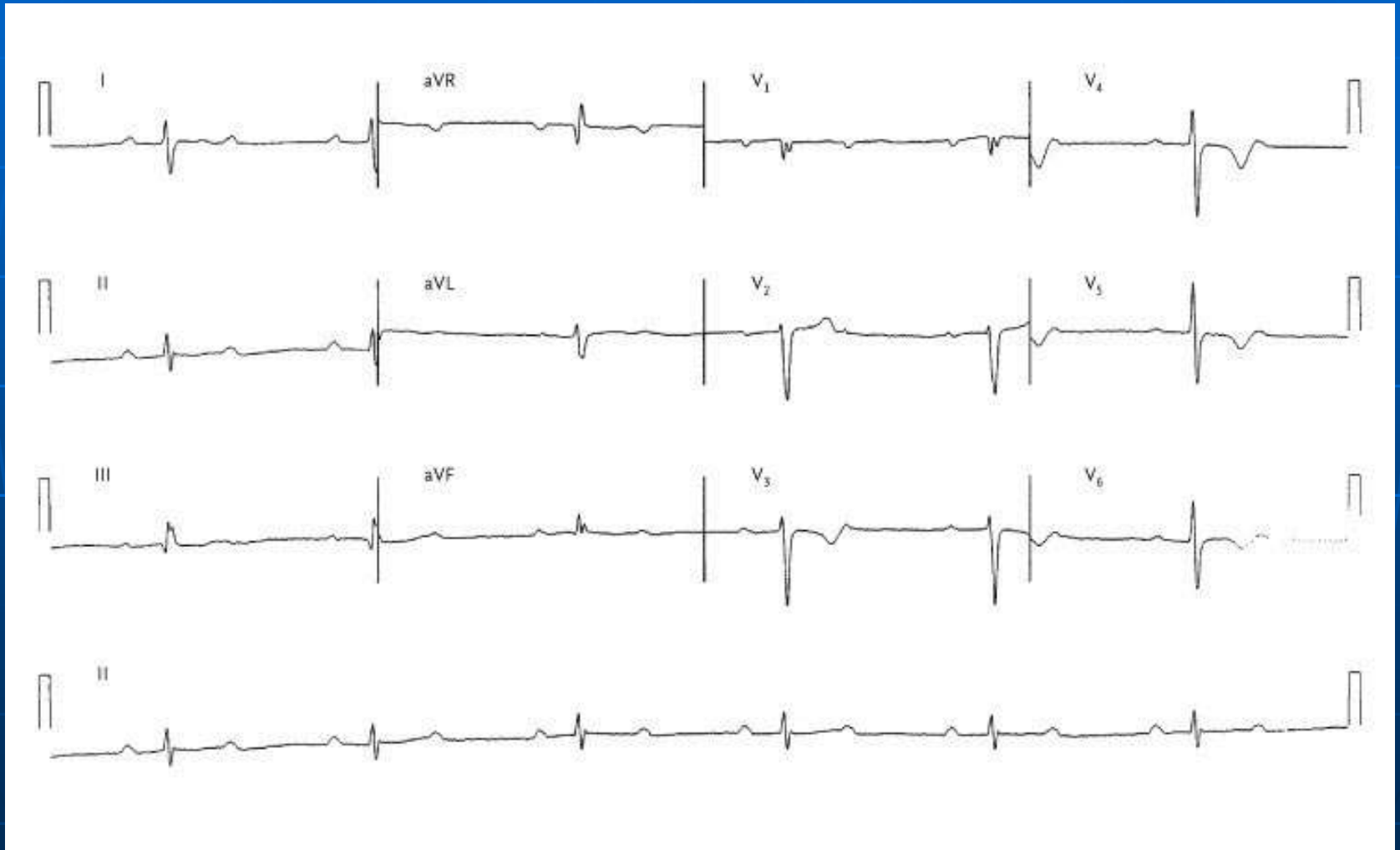
Severity of myocarditis: mild

- mostly focal myocarditis
- **without** heart enlargement
- **without** heart failure – may be LV dysfunction (HF 0-1 stage)
- **no** life-threatening cardiac arrhythmias and conduction disorders
- only nonspecific ECG changes (nonspecific ST-T abnormalities) and tachycardia.

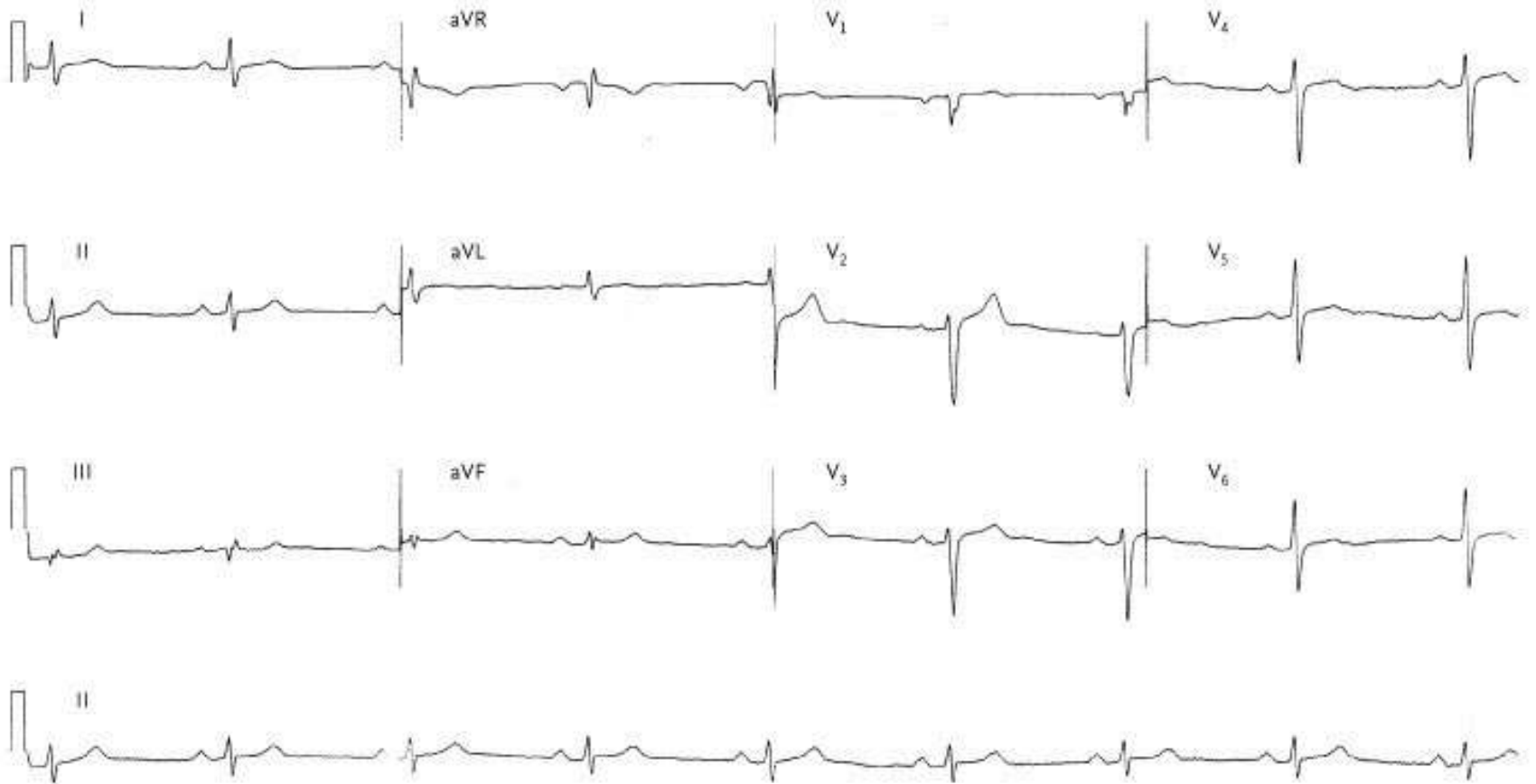
Severity of myocarditis: moderate

- focal or diffuse myocarditis
- enlargement of heart cavities (relative valvular insufficiency)
- moderate heart failure (stage I-IIA)
- involvement of pericardium (myopericarditis)
- no life-threatening cardiac arrhythmias and conduction disorders

Myocarditis (1): AV block 2:1; T-wave inversion



Myocarditis (2) : the same patient after treatment



Myopericarditis

Clinical Features

- Most common-sudden or gradual onset of sharp or stabbing pain with radiation to back, neck, L shoulder or arm
- Radiation to L trapezial ridge is distinguishing
- Pain more severe with lying supine and relieved with sitting
- Low grade fever, dyspnea and dysphagia
- Transient, intermittent friction rub

ECG changes in myopericarditis

EKG-changes in four stages

1-ST elevation in I, V5 and V6, PR depression in II, aVF and V4-V6

2-ST segment normalizes, T wave decreases

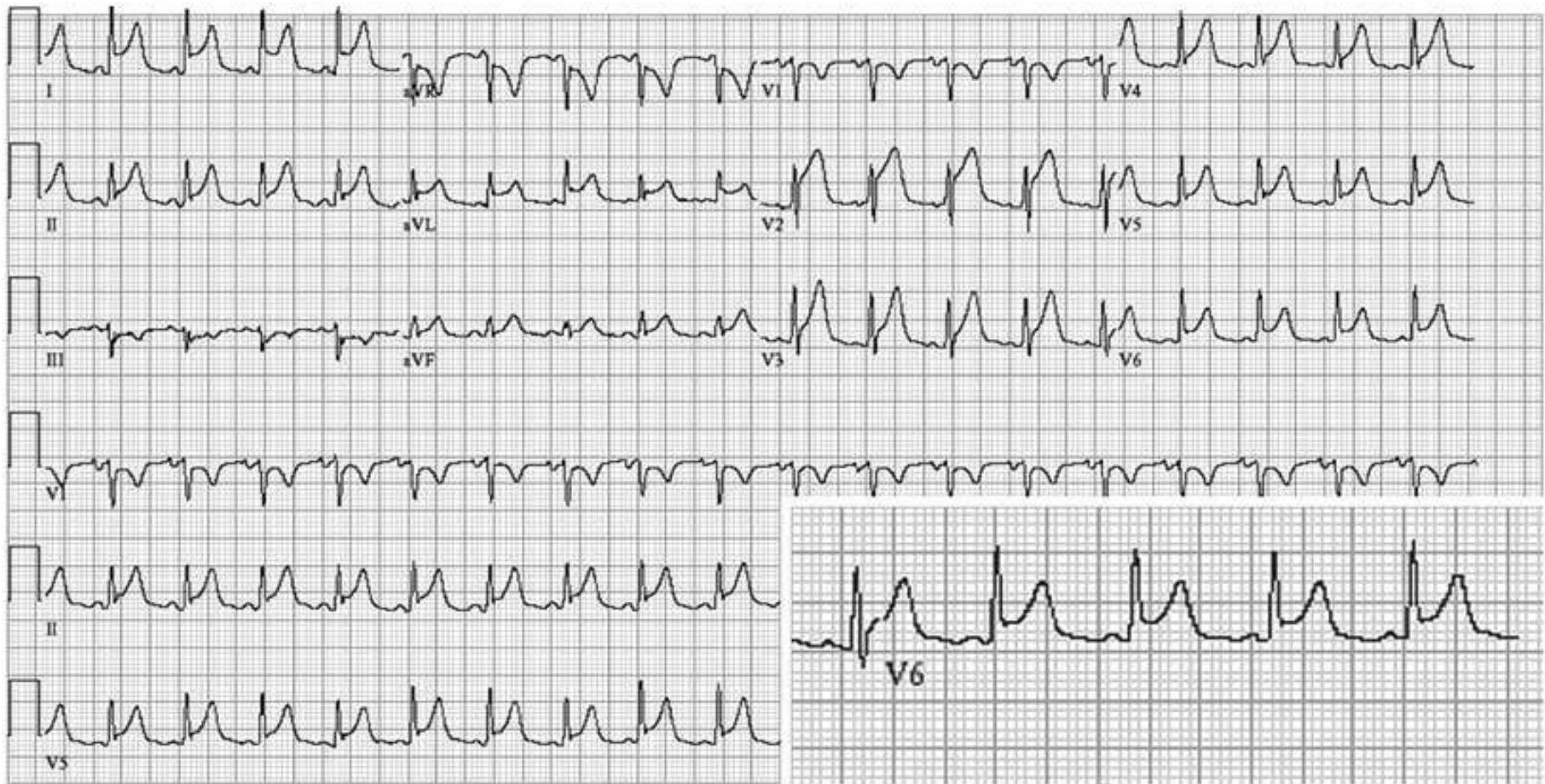
3-Inverted T waves in leads with previous ST elevation

4-Return to normal ECG

In I, V5, or V6 ST:Twave ratio >0.25 most likely acute pericarditis

PQ-depression

Myocarditis with pericardium involvement (myopericarditis): ST-segment elevation and PQ-segment depression



Severity of myocarditis: severe

- predominantly **diffuse** myocarditis with **cardiomegaly**, **significant** heart failure (II A-B stage) with **life-threatening** disorders of cardiac rhythm and conduction.

Natural History of Acute (Viral) Myocarditis

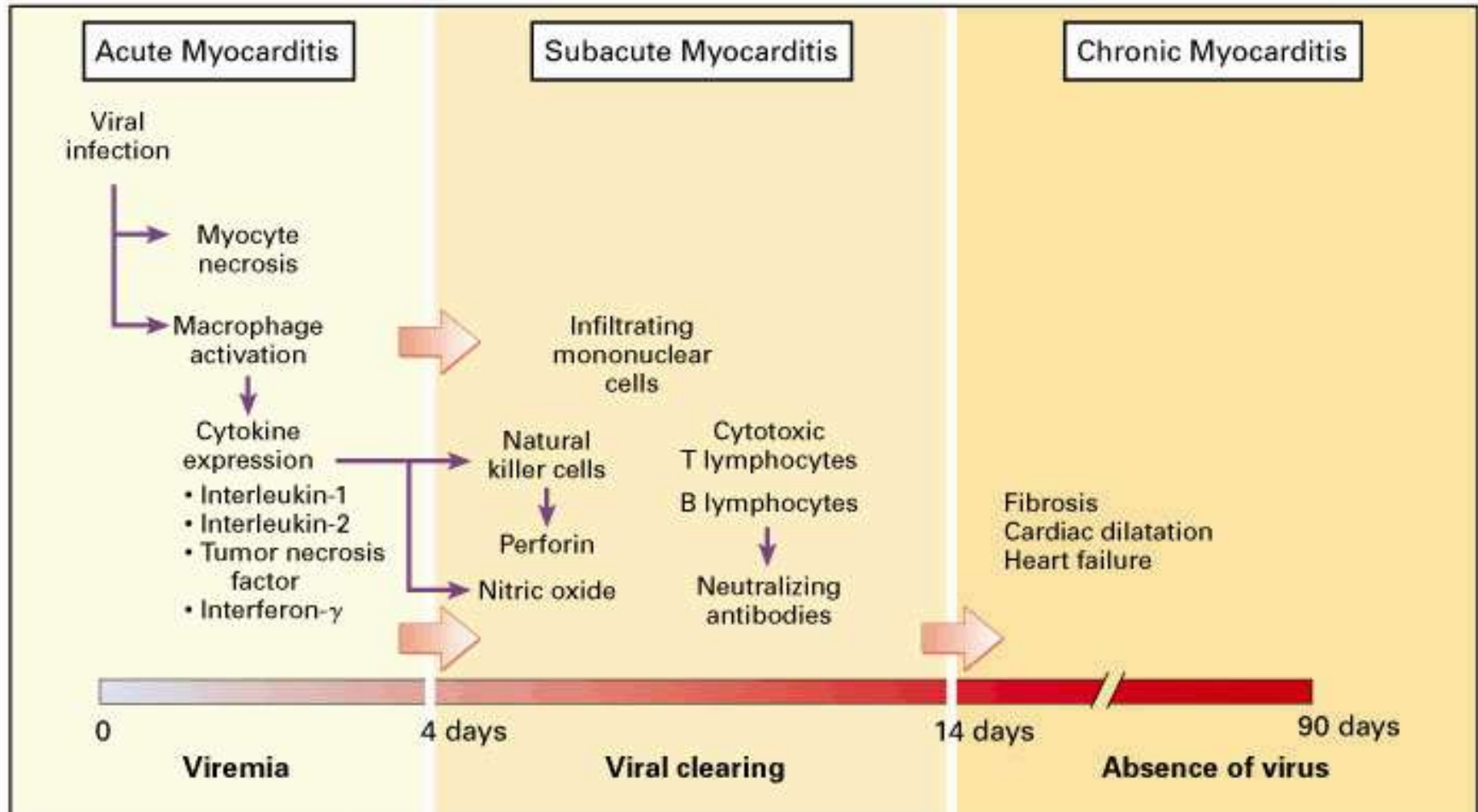
Subclinical, no sequelae

Fulminant; cardiac dilatation, heart failure, arrhythmias, death

Self limited cardiac dysfunction with resolution in weeks/months

Chronic cardiomyopathy

Clinical course and Natural History of Acute (Viral) Myocarditis



Classification of myocarditis : Stage

- 1) acute
- 2) subacute
- 3) chronic
- 4) myocardiofibrosis

Classification of myocarditis :

Etiology

- 1) with established etiology
(infectious, bacterial, viral,
parasitical, other diseases)
- 2) with unspecified etiology

Classification of myocarditis : Morphological features of infiltration (data EMB)

- 1) lymphocytic;
- 2) eosinophilic;
- 3) giant cell;
- 4) granulomatous

Classification of myocarditis : Spreading

- 1) focal
- 2) diffuse

Classification of myocarditis : severity

- 1) mild
- 2) moderate
- 3) severe

Classification of myocarditis

VI. **Complications:** Cardiac rhythm and conduction abnormalities, thromboembolism, and others.

VII. **Heart failure:** stage 0-III, I-IV FC.

Treatment: etiotropic treatment

- **Antibiotic therapy** – only after the confirmation of the etiological factor (diphtheria)
- **Antiviral drugs** – only for **proven** viral myocarditis (etiologic agent is known AND sensitive to antiviral drug)
- However, beneficial effects are seen only if therapy is started **prior to** inoculation or **soon thereafter**.
- Nevertheless, antiviral therapy may be considered in acute, fulminant myocarditis, in institutional outbreaks and in laboratory-acquired cases

Treatment: pathogenetic therapy

Avoidance of exercise

- Physical activity should be restricted to reduce the work of the heart during the acute phase of myocarditis, especially when there is fever, active systemic infection, or heart failure

Terms of physical activity limitation

- 10 – 14 days – mild myocarditis (up to ECG normalization)
- 4 – 6 weeks – moderate myocarditis (up to normalization of heart size)
- Individually – severe myocarditis (up to decreasing of HF severity and disappearance of rhythm disorders)

Treatment: symptomatic

- Treatment of systolic heart failure (ACEI/ARA, beta-blockers, diuretics, aldosterone antagonists, digoxin in atrial fibrillation/flutter)
- Treatment of arrhythmias (beta-blockers, amiodarone)
- Anticoagulation to prevent thromboemboli (mural thrombi) – warfarin (INR 2,0 – 3,0)
- NSAID – only for **perimyocarditis** with **normal** LV function and prominent chest **pain** from pericarditis.
- Pacemaker for complete AV block
- ? prednisone and azathioprine - **no** apparent benefit seen in the Myocarditis Treatment Trial

Cardiomyopathies

Cardiomyopathy

- The term 'cardiomyopathy' was first used in 1957 by Brigden, who described a group of uncommon, non-coronary myocardial diseases
- In 1961 Goodwin defined cardiomyopathies as 'myocardial diseases of unknown **cause**'. He described three different entities, namely 'dilated, hypertrophic and restrictive', terms which **are still in use** today
- Controversial classifications

Cardiomyopathy Classification

Classification (2 systems)

- **Based on Suspected Etiology**
 - **Primary** -defined as those involving only the heart
 - **may be genetic, mixed (genetic or non genetic), or acquired**
 - **Secondary** -characterized by a generalized multiorgan involvement
 - **which are accompanied by other organ system involvement**

World Health Organization and International Society and Federation of Cardiology classify on the basis of their pathologic or pathophysiologic features (more accepted system) – All are based off of echocardiography - 1980 year 3 types:

- **Dilated** – usually mostly Systolic dysfunction
- **Restrictive** – usually mostly Diastolic dysfunction
- **Hypertrophic** – usually mostly Diastolic dysfunction

Cardiomyopathy Classification

In 1996 World Health Organization and International Society and Federation of Cardiology added:

- arrhythmogenic right ventricular dysplasia" (with the inappropriate term ``dysplasia" later changed to ``cardiomyopathy")
- a group of ``unclassified cardiomyopathies", defined as ``those that do not fit in any group"

Definition of the cardiomyopathies

European Society of Cardiology, 2008

“myocardial disorders in which the heart muscle is structurally and functionally abnormal in the absence of coronary artery disease, hypertension, valvular or congenital heart disease sufficient to cause the observed myocardial abnormality”

Groups of the cardiomyopathies

European Society of Cardiology, 2008

1. Hypertrophic
2. Dilated
3. Restrictive
4. Arrhythmogenic right ventricular cardiomyopathy
5. Unclassified
 - LV non-compaction
 - Takotsubo cardiomyopathy

Cardiomyopathy

- 3rd most common form of heart disease in U.S.
- 2nd most common cause of adolescent sudden death (hypertrophic obstructive cardiomyopathy)

Hypertrophic cardiomyopathy (HCM)

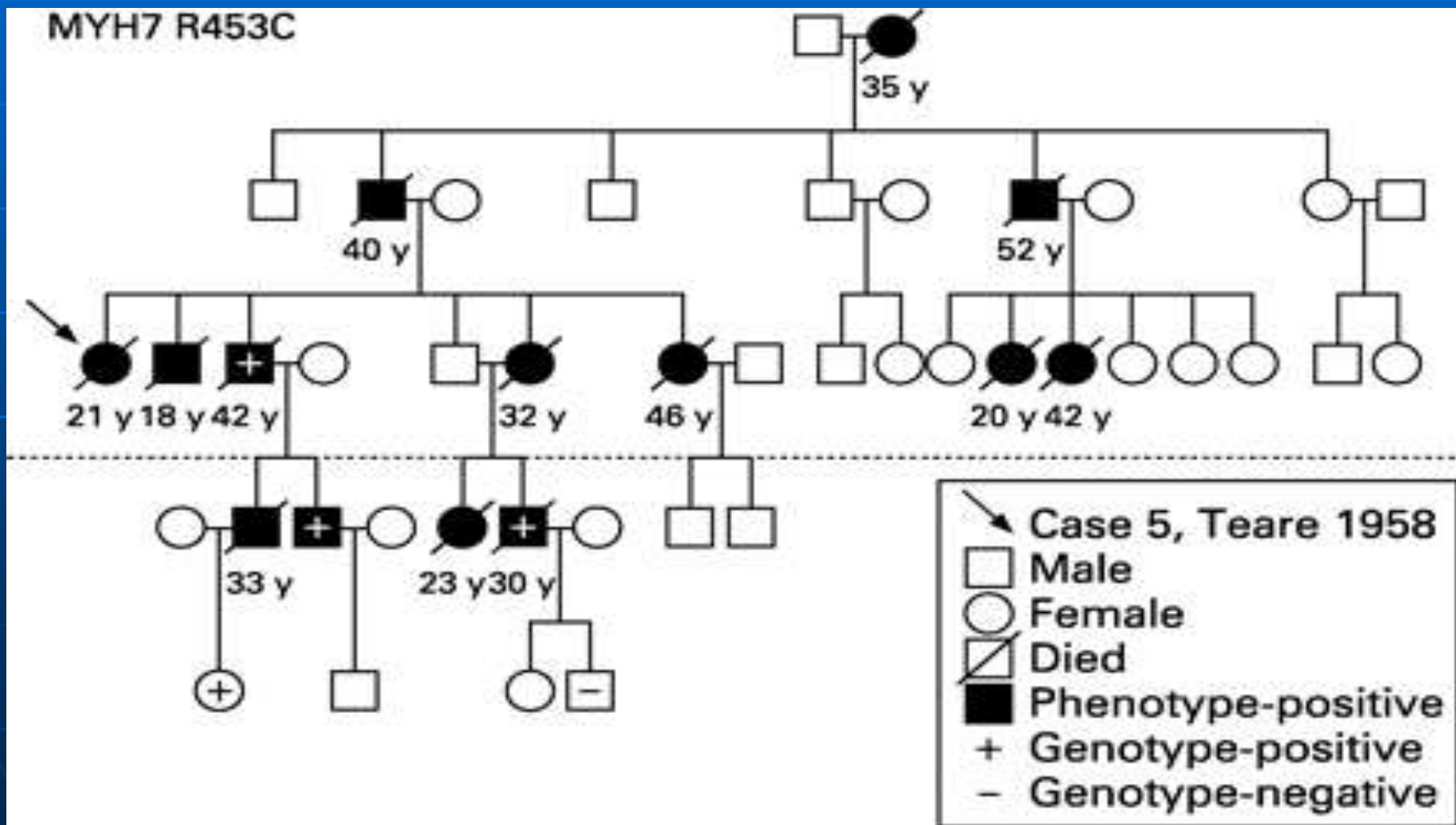
- HCM is a genetic disease characterized by unexplained LV hypertrophy, associated with non-dilated ventricular chambers, in the absence of another cardiac or systemic disease capable of producing that degree of hypertrophy

Hypertrophic Cardiomyopathy

- prevalence of hypertrophic cardiomyopathy in the absence of aortic valve disease or systemic hypertension is at least 1:500 of the adult population
- Mortality 1%, 4-6% in childhood/adolescence
- Sarcomeric gene mutations are the most frequent cause of HCM, accounting for approximately 30 - 65% of cases
- A small proportion of patients with the HCM phenotype are affected by neuromuscular diseases (e.g. Frederich's ataxia), mitochondrial diseases, metabolic disorders or lysosomal storage diseases (i.e. Fabry, Pompe, Danon)

Pedigree of a family with HCM which was described for the 1st time in 1957-1958yy.

Brock RC. Functional obstruction of the left ventricle. *Guy's Hosp Rep* 1957;106:221-38; Teare D. Asymmetrical hypertrophy of the heart in young adults. *Br Heart J* 1958;20:1-18.



Hypertrophic cardiomyopathy; diagnostic criteria

- HCM is diagnosed by a maximal LV wall thickness greater than **15 mm**, based on echocardiography (ECHO) or cardiac magnetic resonance (CMR).
- This value is lowered to 13 - 14 mm, when family members are screened.
- Abnormal compliance-impaired diastolic relaxation and filling-output usually normal

HCM: Echocardiographic and cardiac magnetic resonance images from a 17-year old female patient



Parasternal long and short axis views show severe LV thickness values with redundant mitral leaflets and small cavity size.

HCM: Echocardiographic and cardiac magnetic resonance images from a 17-year old female patient



Apical 4 chambers view shows massive hypertrophy of the septum and the anterolateral wall

The distribution of hypertrophy is usually asymmetric and sometimes confined to one or two LV segments. As a consequence, LV mass (measured by CMR) can be within the normal range. LV outflow tract obstruction is an important feature of HCM, and may be demonstrated in up to 70% patients



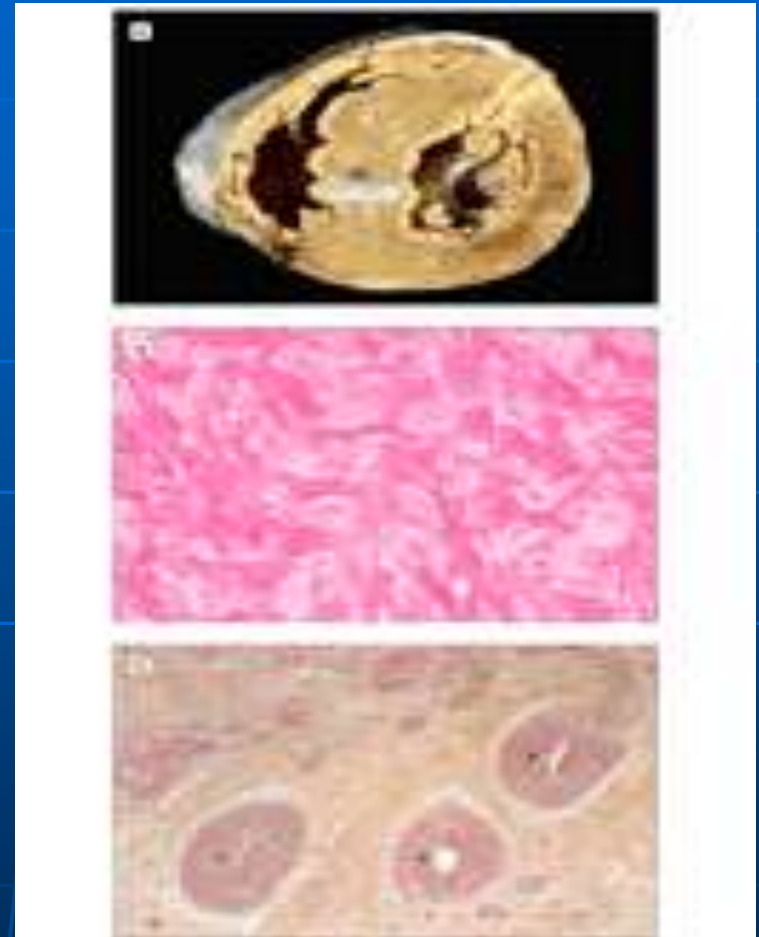
HCM

A. Massive hypertrophy of the septum

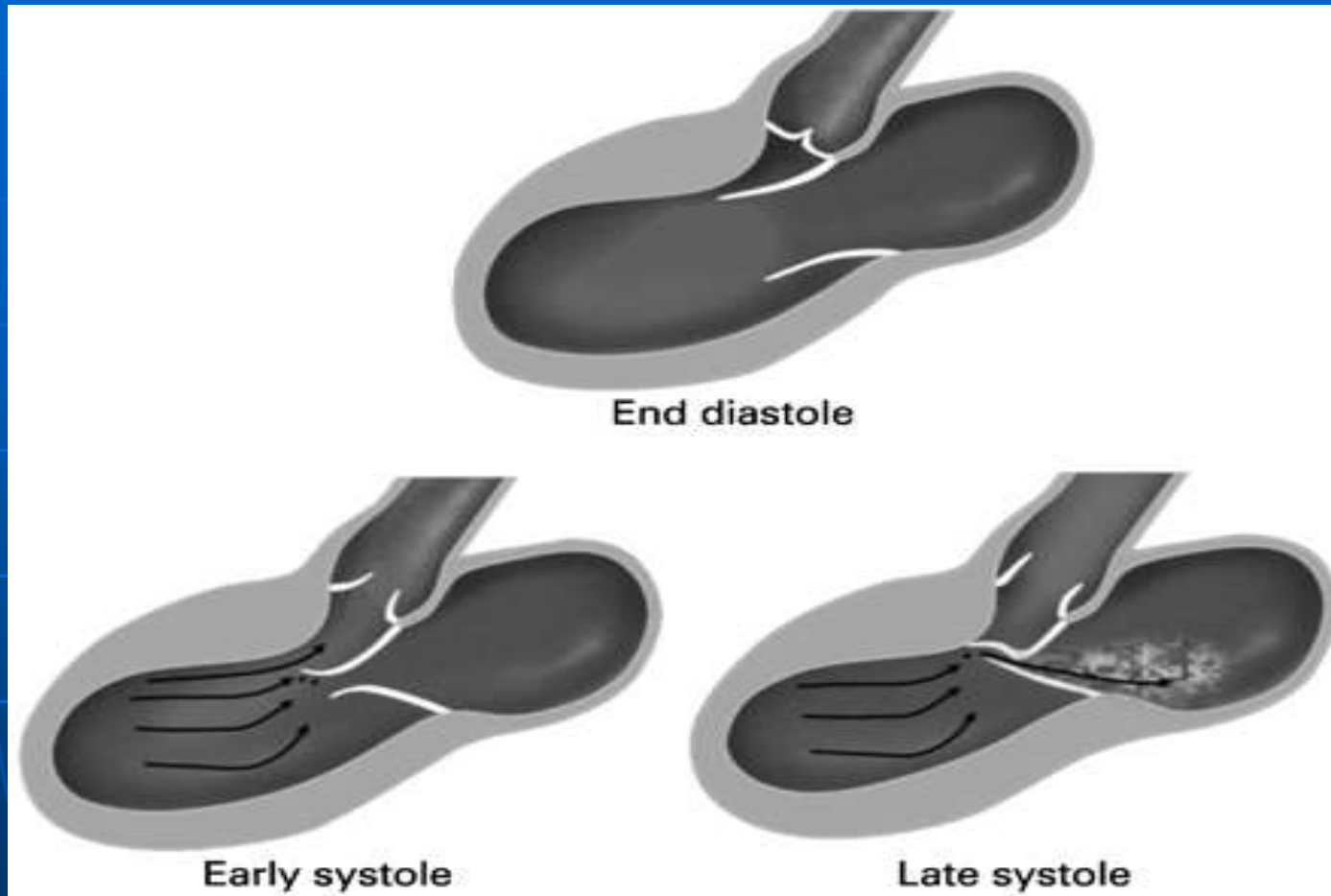
B. Chaotic arrangement of of cardiomyocytes

C. Wall thickening and narrowing of the coronary arteries

(hematoxylin and eosin x50). Adapted from Maron BJ. Hypertrophic cardiomyopathy. *Current Probl Cardiol.* 1993;18:637-704



Left ventricular outflow obstruction in HCM



In early systole (bottom left), abnormal flow around the hypertrophied septum pushes the mitral valve into the outflow tract and results in obstruction and mitral valve regurgitation (bottom right).

Hypertrophic Cardiomyopathy

■ Presentation (HF diastolic, ischemia, arrhythmia)

- Dyspnea on exertion,
- ischemic chest pain
- Palpitations, arrhythmias, syncope, pre-syncope
- Sudden Death (usually due to V-tach during exercise)

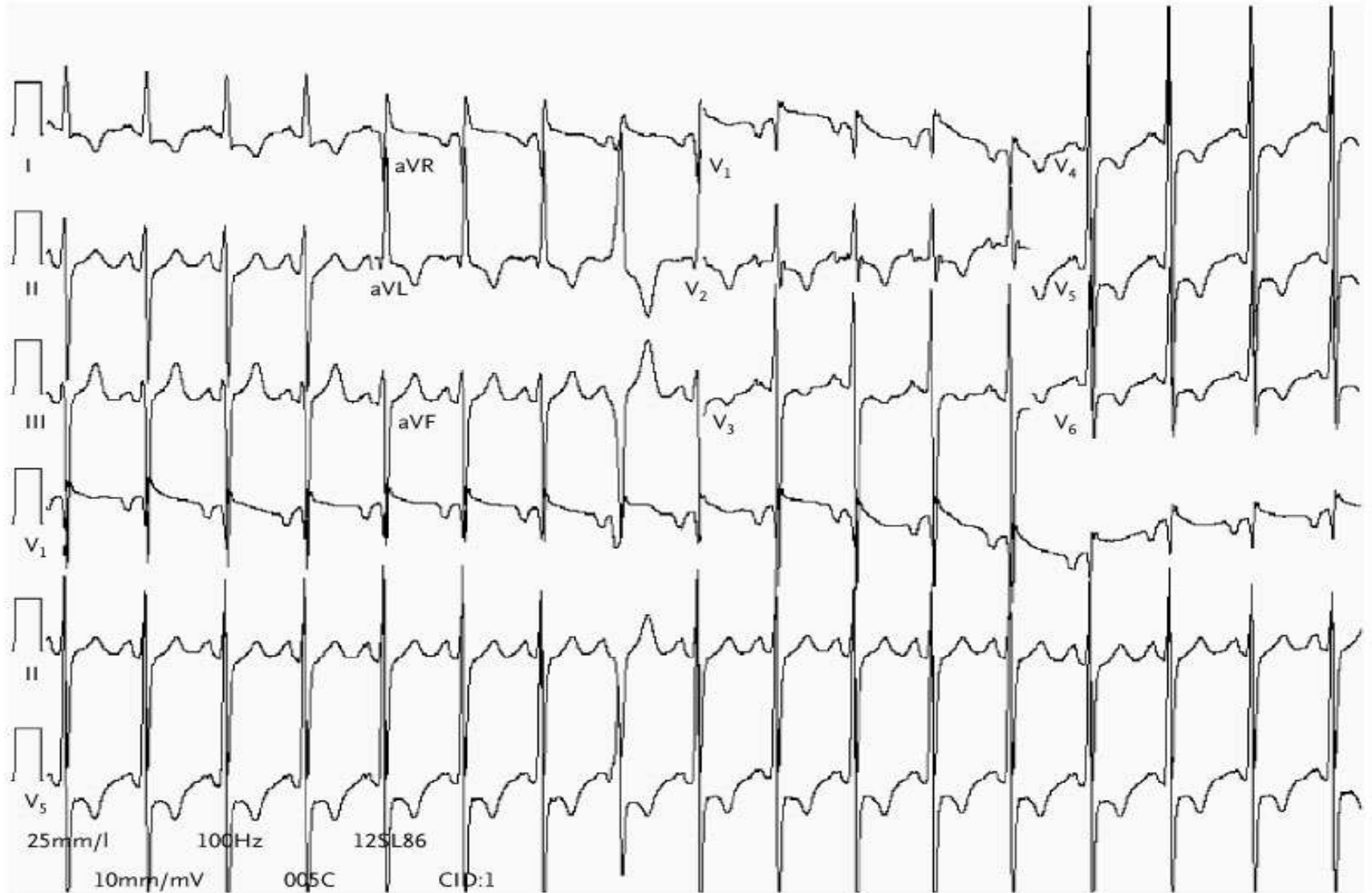
■ Physical Exam

- S4 gallop
- prominent systolic ejection murmur at left sternal border that increases with valsalva, sudden standing, or exercise – in the presence of LV outflow obstruction)

Hypertrophic Cardiomyopathy

- Diagnosis
 - Chest Xray: usually normal
 - Echocardiogram: LVH with disproportionate septal hypertrophy; small ventricular volume
 - ECG: LVH (30%) , & LAH (25-50%); Large septal Q waves-25%, Afib and PVCs common
- * think hypertrophic cardiomyopathy in any young person whose EKG shows large septal Q waves*

ECG in HCM



Hypertrophic Cardiomyopathy

■ Treatment

- Can become unstable in afib due to lack of atrial kick (cardioversion & heparinization)
- Long term care with **Beta blockers**
- Amiodarone is treatment of choice for ventricular dysrhythmias
- Diuretics in the face of pulmonary congestion
- Avoid agents that reduce ventricular volume (nitrates) or increase myocardial contractility (digoxin)
- Antibiotic prophylaxis for dental procedures
- Avoidance of competitive athletics
- Anticoagulation if in afib

Dilated cardiomyopathy (DCM)

DCM is characterized by LV dilatation and global systolic dysfunction (EF < 50%), in the absence of coronary artery disease or other identifiable causes (such as systemic hypertension, valve disease, drugs, inflammatory heart diseases) capable of causing that magnitude of impairment

Dilated Cardiomyopathy

- incidence - 5 to 8 cases per 100,000 population
- prevalence of 36 per 100,000
- African Americans and males have 2.5x increased risk
- Most common age of diagnosis 20-50yrs
- 80% of DCM cases are idiopathic

Dilated Cardiomyopathy

Clinical Presentation: symptoms

Usually presents as unexplained heart failure (prior to echo)

- Symptoms of CHF-dyspnea on exertion, orthopnea and paroxysmal nocturnal dyspnoe.
- Chest pain can occur due to low coronary vascular reserve
- Palpitations

Dilated Cardiomyopathy

Clinical Presentation: Signs

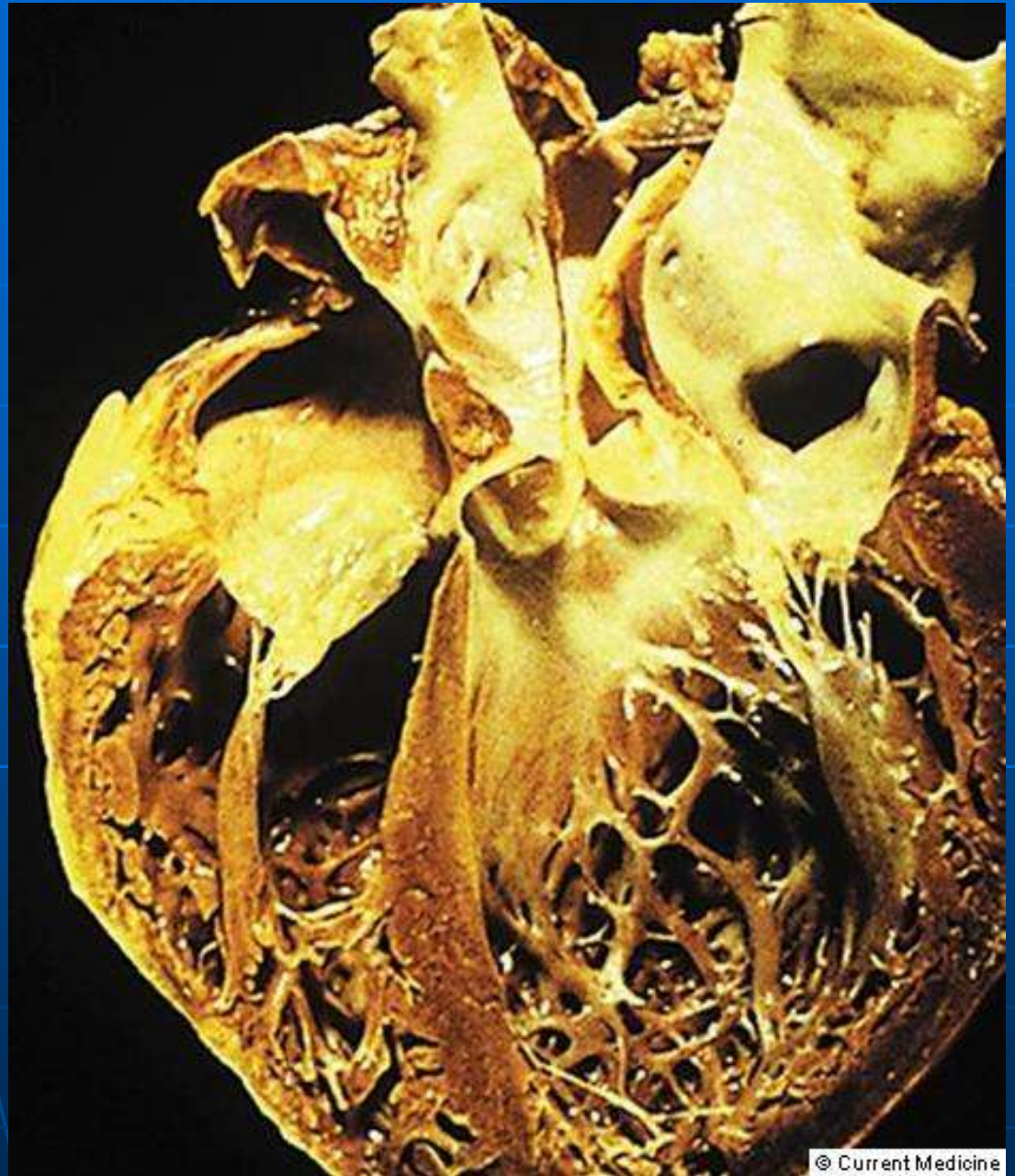
- Rales, S3 S4 gallops, narrow pulse pressure, murmurs of mitral or tricuspid regurgitation
- Manifestations of embolization neurologic deficits, flank pain, hematuria, pulseless cyanotic extremity

Dilated Cardiomyopathy

Diagnosis

- Holosystolic regurgitant murmur or gallop may be present
- Dependent edema, bibasilar rales
- CXR- enlarged heart, biventricular enlargement, and pulmonary vascular congestion
- ECG- LVH, Left atrial enlargement, Q waves, poor R wave progression, afib
- **Echo-Confirms Dx.**- Dilation of all four chambers (ventricles greater than atria); increased muscle mass; systolic failure - increased systolic and diastolic volumes, decreased EF, valvular regurgitation (relative valvular insufficiency); mural thrombi

Heart chambers dilatation in DCM



Dilated Cardiomyopathy

Treatment

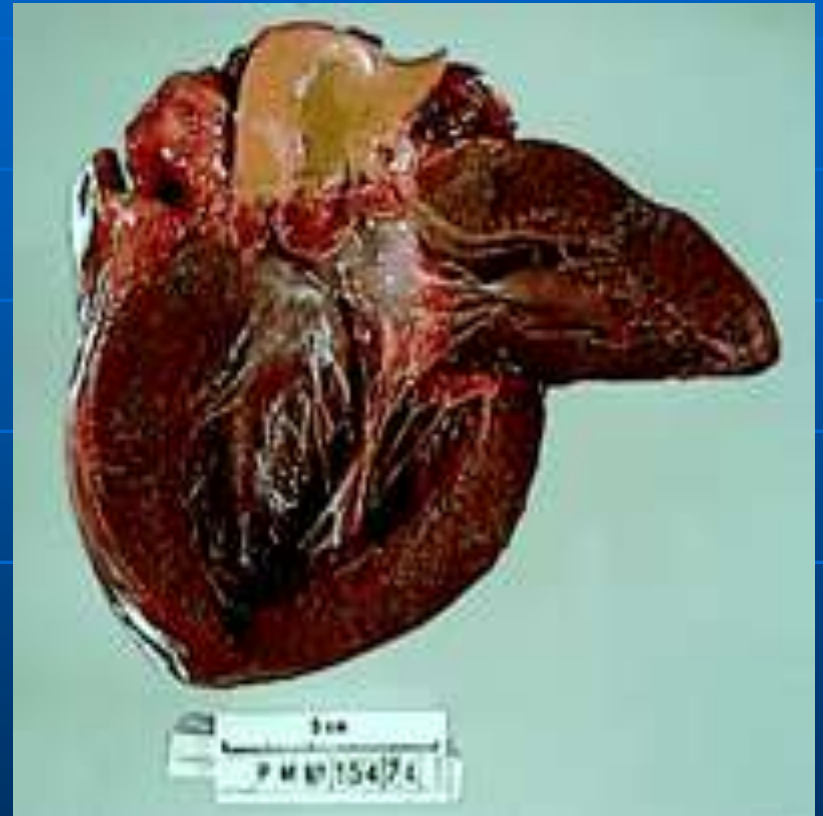
- HF - ACE inhibitors and B-Blockers- improve survival;
- Amiodarone- for complex ventricular ectopy
- Anticoagulation can be considered: all with mural thrombi; evidence of pulmonary or systemic emboli; atrial fibrillation

Restrictive Cardiomyopathy (RCM)

- RCM is defined by the presence of a restrictive LV physiology, with normal or more often reduced diastolic/systolic volumes, normal wall thickness and systolic function, marked diastolic flow impairment and **bi**atrial dilatation.

Restrictive Cardiomyopathy (amyloidosis)

- LV normal wall thickness and small cavities



Restrictive Cardiomyopathy

- RCM are rather uncommon, although their prevalence is still unknown.
- Mostly idiopathic- sometimes familial
- Systemic disorders-amyloidosis, sarcoidosis, hemochromatosis, scleroderma, and carcinoid.

Restrictive Cardiomyopathy

Clinical Features

- Symptoms of CHF-dyspnea, orthopnea, pedal edema- rare chest pain
- Exam-may have S3 or S4 gallop, regurgitation murmurs, rales, jugular vein distension, Kussmaul's sign (jvd with inspiration), hepatomegaly, pedal edema or ascites

Restrictive Cardiomyopathy

Diagnosis

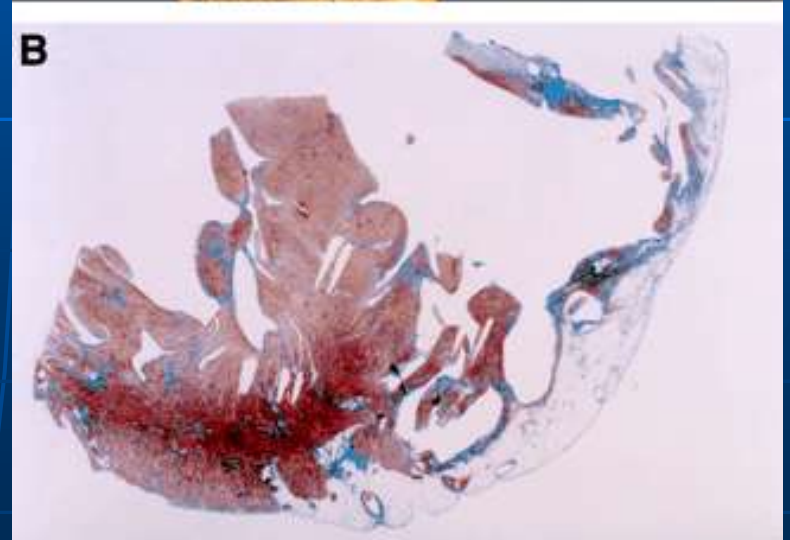
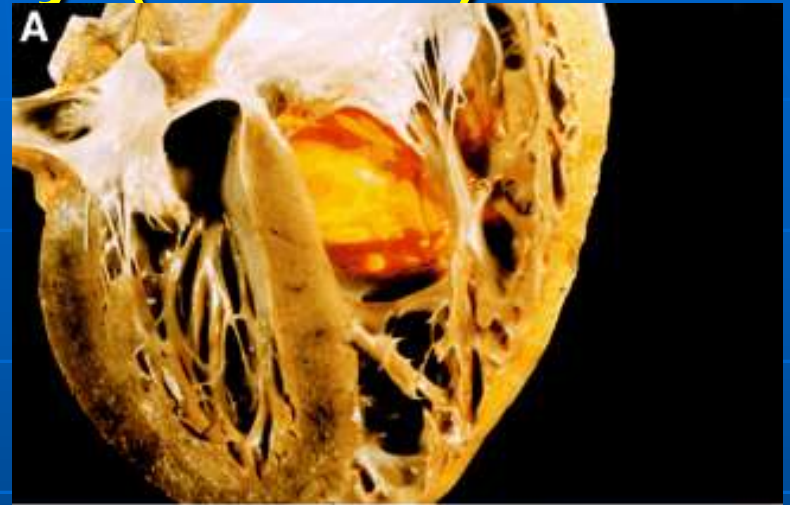
- CXR-signs of CHF without cardiomegaly
- ECG-nonspecific changes most likely
- **Conduction** disturbances and low-voltage **QRS** complexes are common with amyloidosis or sarcoidosis
- Ultrasound - Low end diastolic volume; Decreased cardiac output; markedly dilated atria, normal systolic function, mitral/tricuspid regurgitation

Restrictive Cardiomyopathy: Treatment

- Symptom directed-diuretics and ACE inhibitors
- Corticosteroids for sarcoidosis
- Chelation therapy for hematochromatosis

Arhythmogenic Right Ventricular Cardiomyopathy (ARVC)

- ARVC is characterized by fibrofatty replacement of the right ventricular myocardium and ventricular arrhythmias

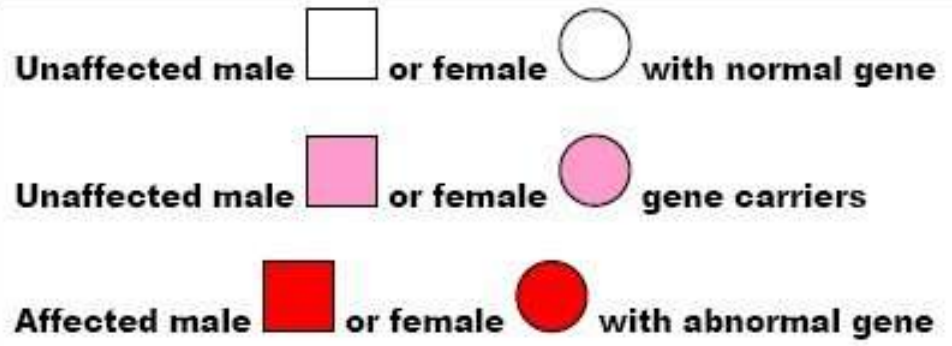
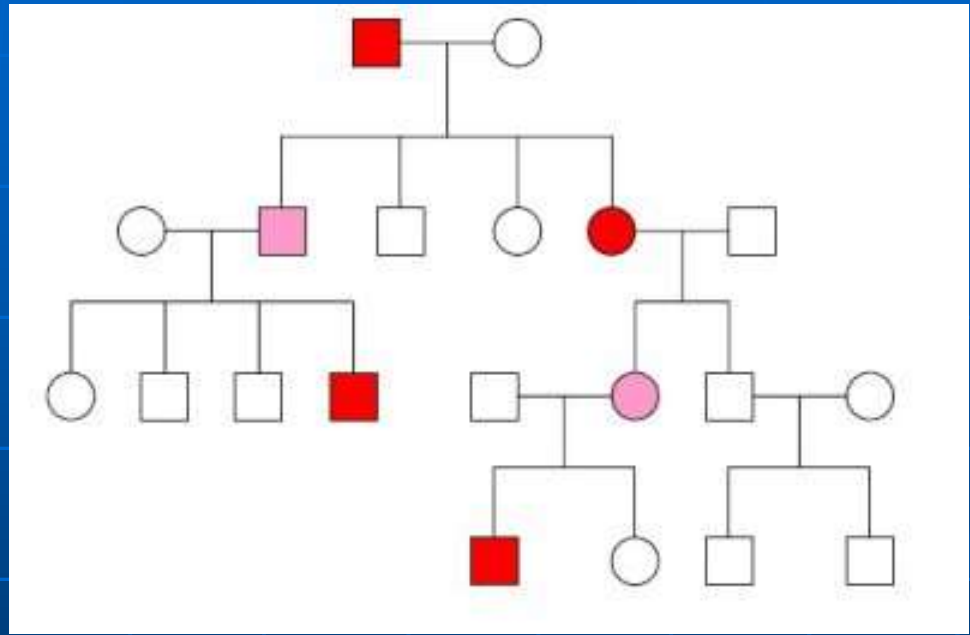


Arhythmogenic Right Ventricular Cardiomyopathy (ARVC)

- Most rare form of cardiomyopathy
- **Ventricular arrhythmias** are the clinical hallmark of the disease, but atrial fibrillation may also occur.
- Major cause of SCD in young, especially in some regions of Italy, still exists in US - consider in young pt with syncope, palpitations, and aborted SCD
- Typical presentation of sudden death in young or middle aged pt
- Familial in 50% of cases

Arhythmogenic Right Ventricular Cardiomyopathy (ARVC)

ARVC is generally a familial disease with autosomal dominant inheritance but it may be recessive when associated with woolly hair and palmopalmar hyperkeratosis (eg, Naxos disease, Carvajal syndrome)



Arhythmogenic Right Ventricular Cardiomyopathy (ARVC)

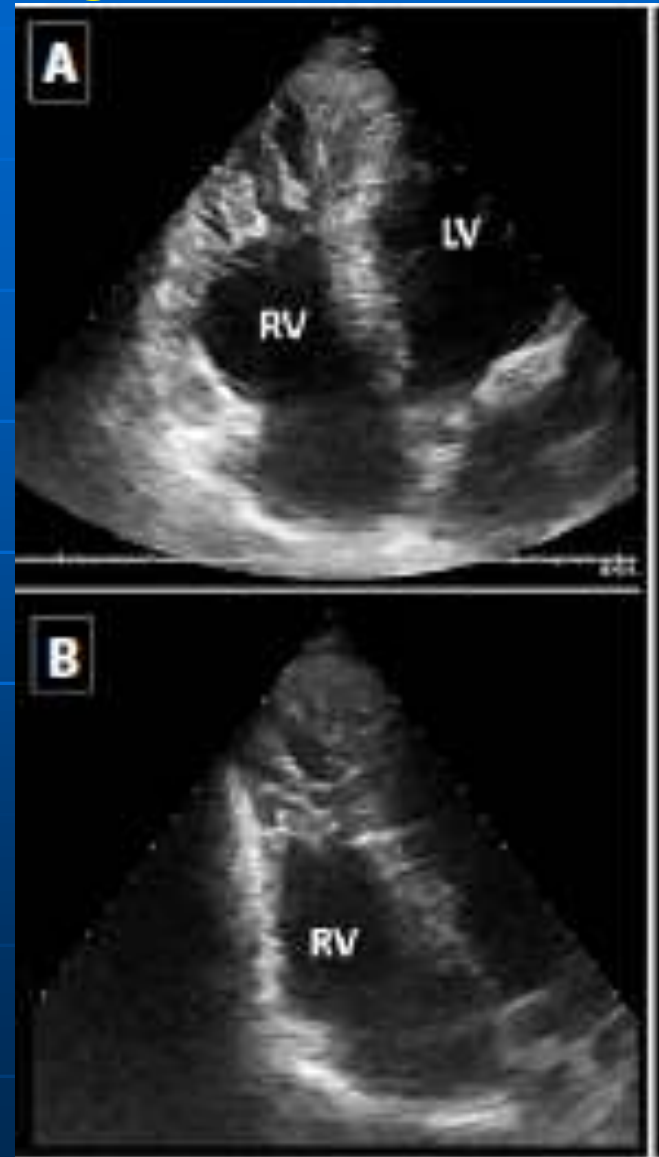
- Exam usually normal
- **ECG**- inverted T waves from V1 to V4, RBBB and isolated premature ventricular beats with an LBBB morphology may be present
- **Echo-necessary for diagnosis:**
 - In the most common right-dominant form, structural changes may be absent or confined to a localized region of the right ventricle (inflow and outflow tract, right ventricular apex, known as the '**triangle of dysplasia**') at an early stage

Twelve lead ECG : inverted T waves from V1 to V4 and isolated premature ventricular beats with an LBBB morphology



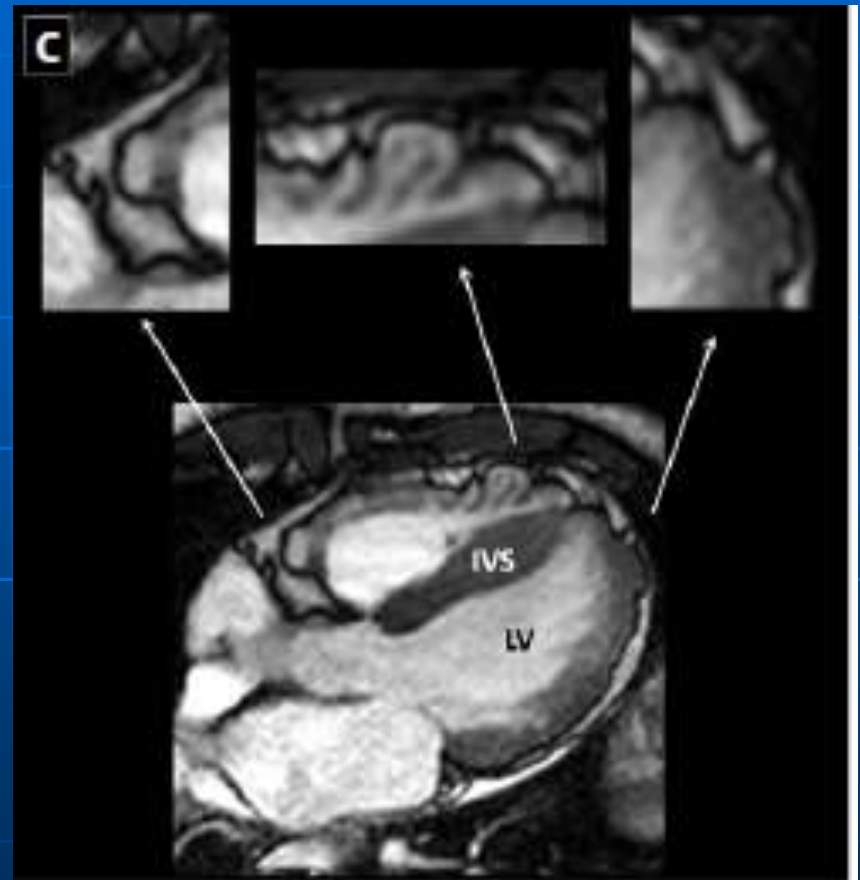
Echocardiographic image of ARVC

evident systolic bulging in infundibular, apical, and subtricuspid regions of the RV (**triangle of dysplasia**)



CMR images of ARVC

wall aneurysms
within the 'triangle
of dysplasia'



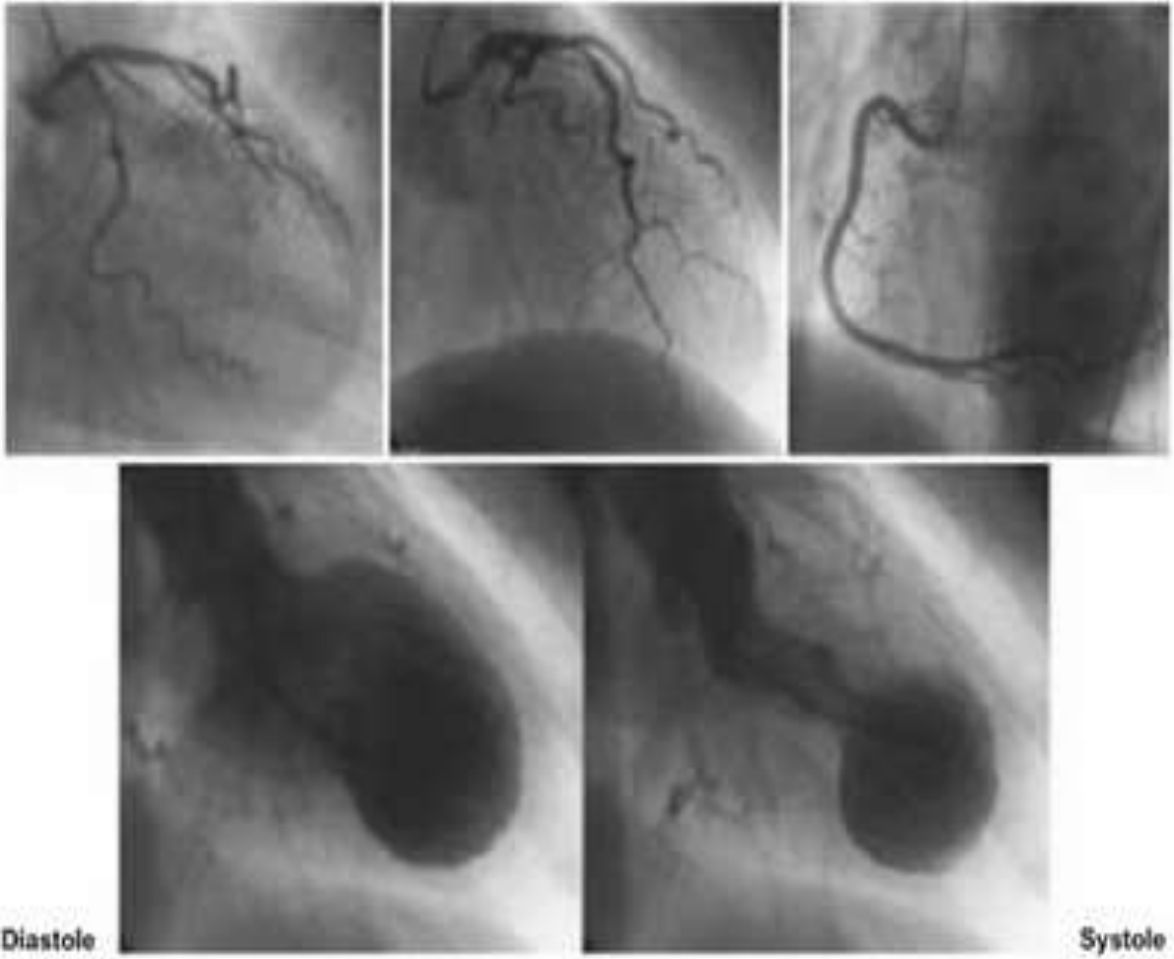
ARVC: Treatment

- β blockers and class I and III antiarrhythmic drugs
- catheter ablation
- implantable cardioverter defibrillator (ICD).

Tako-Tsubo cardiomyopathy

- LV apical ballooning after a high catecholamine stress which results in LV shape similar to octopus pot (takotsubo pot which is Japanese octopus trap)
- Has been described following stressful event like hypoglycemia, earthquakes, following surgery, after emotional stress

Tako-Tsubo cardiomyopathy



Tako-Tsubo cardiomyopathy: clinical features

- Presents as acute anterior MI with chest pain or SOB
- Usually in post-menopausal women
- Cardiac catheterization reveals clean coronary arteries
- Prognosis is good unless there is serious complication (like MR, ventricular rupture, v-tachycardia)

Tako-Tsubo cardiomyopathy: treatment

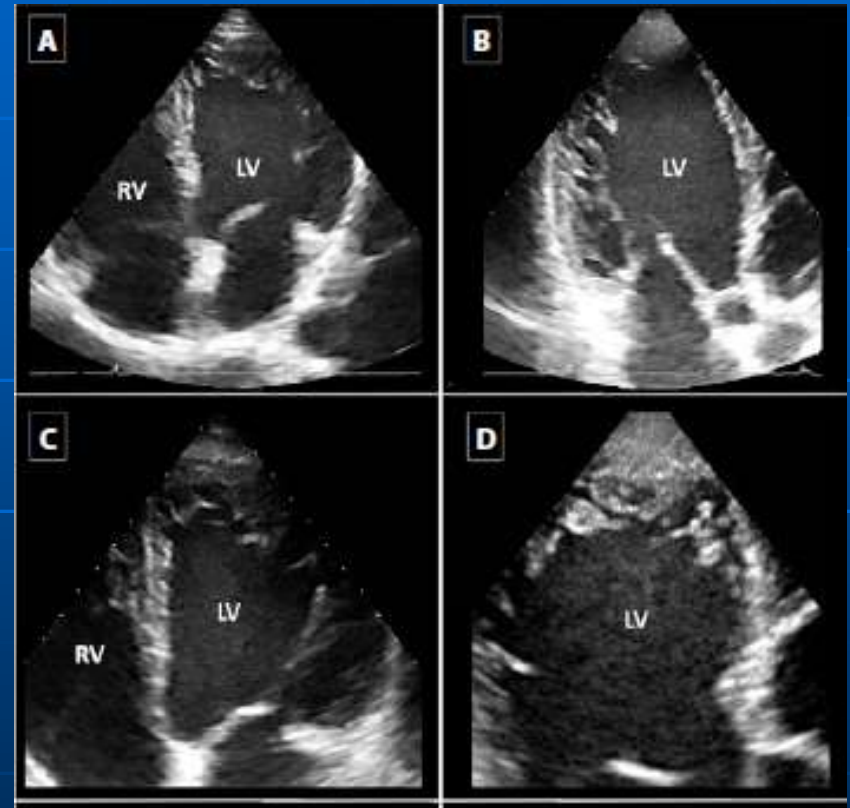
- Preventing of excessive sympathetic activation by combining alpha and beta blockade.
- Beta blockers are used to treat dynamic left ventricular obstruction.
- Phenylephrine may represent an alternative approach in patients presenting with outflow tract obstruction and severe hypotension.
- In hemodynamically unstable patients, early administration of intra-aortic balloon pump counterpulsation should be considered

Isolated LV non-compaction (LVNC)

- is characterized by prominent LV trabeculae and deep inter-trabecular recesses, that can be associated with LV dilatation and systolic dysfunction
- Congenital disorder
- Can be isolated or occur with other congenital heart diseases
- Facial abnormalities and neurologic problems also occur in high proportion of pts with LVNC
- Some genetic links, screen 1st degree relatives

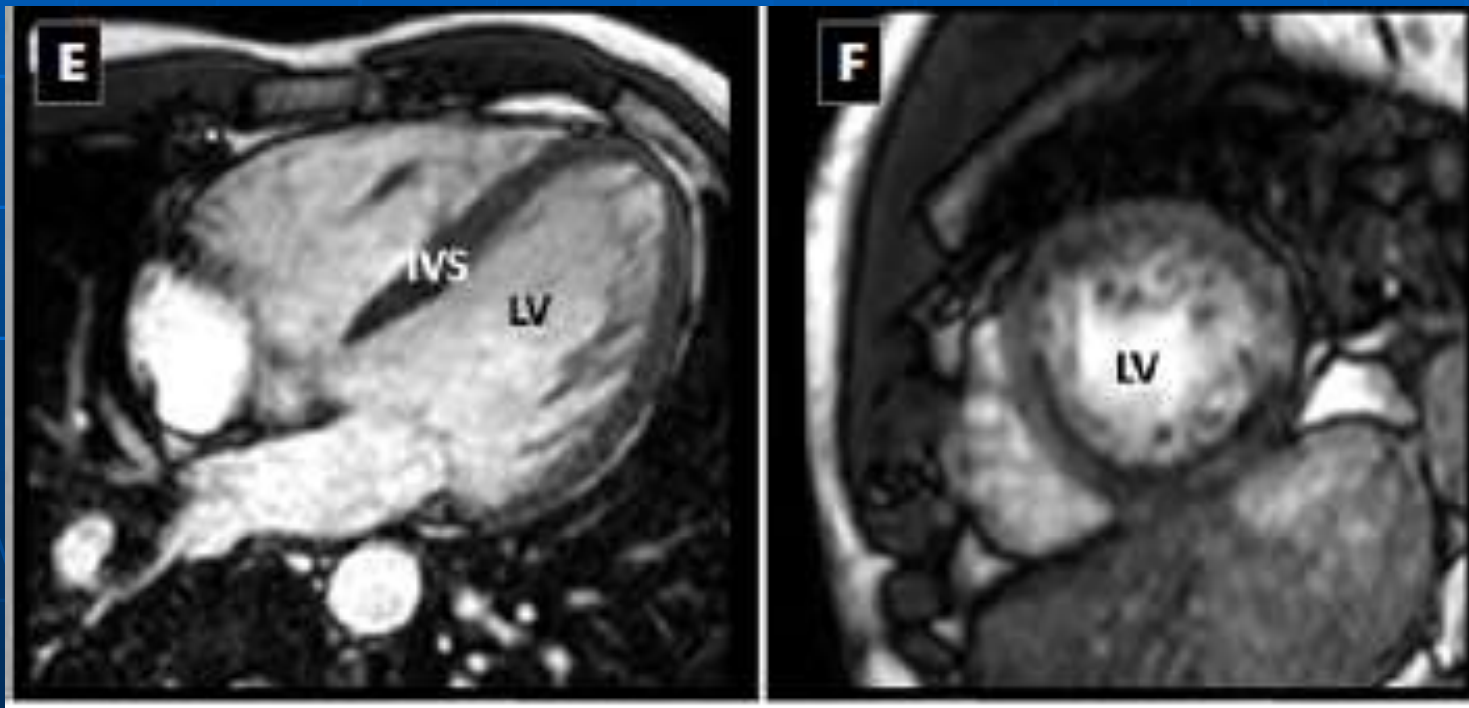
Isolated LV non-compaction (LVNC)

- Multiple trabeculations and recesses are evident, particularly in the apex and the free wall of the LV



Isolated LV non-compaction (LVNC)

- CMR confirmed the diagnosis: Multiple trabeculations and recesses



Treatment of LVNC

Treatment of:

- HF
- Rhythm abnormalities