CARDIOMYOPATHY

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CARDIOMYOPATHY

- ***Cardiomyopathy - heart disease resulting from an abnormality in the myocardium***
- ***Diseases of the myocardium usually produce abnormalities***

  - in cardiac wall thickness

  - chamber size

  - mechanical and/or electrical dysfunction,

CARDIOMYOPATHY

- ***Primary cardiomyopathies** are diseases predominantly confined to the heart muscle*

- ***secondary cardiomyopathies** have myocardial involvement as a component of a systemic or multiorgan disorder*

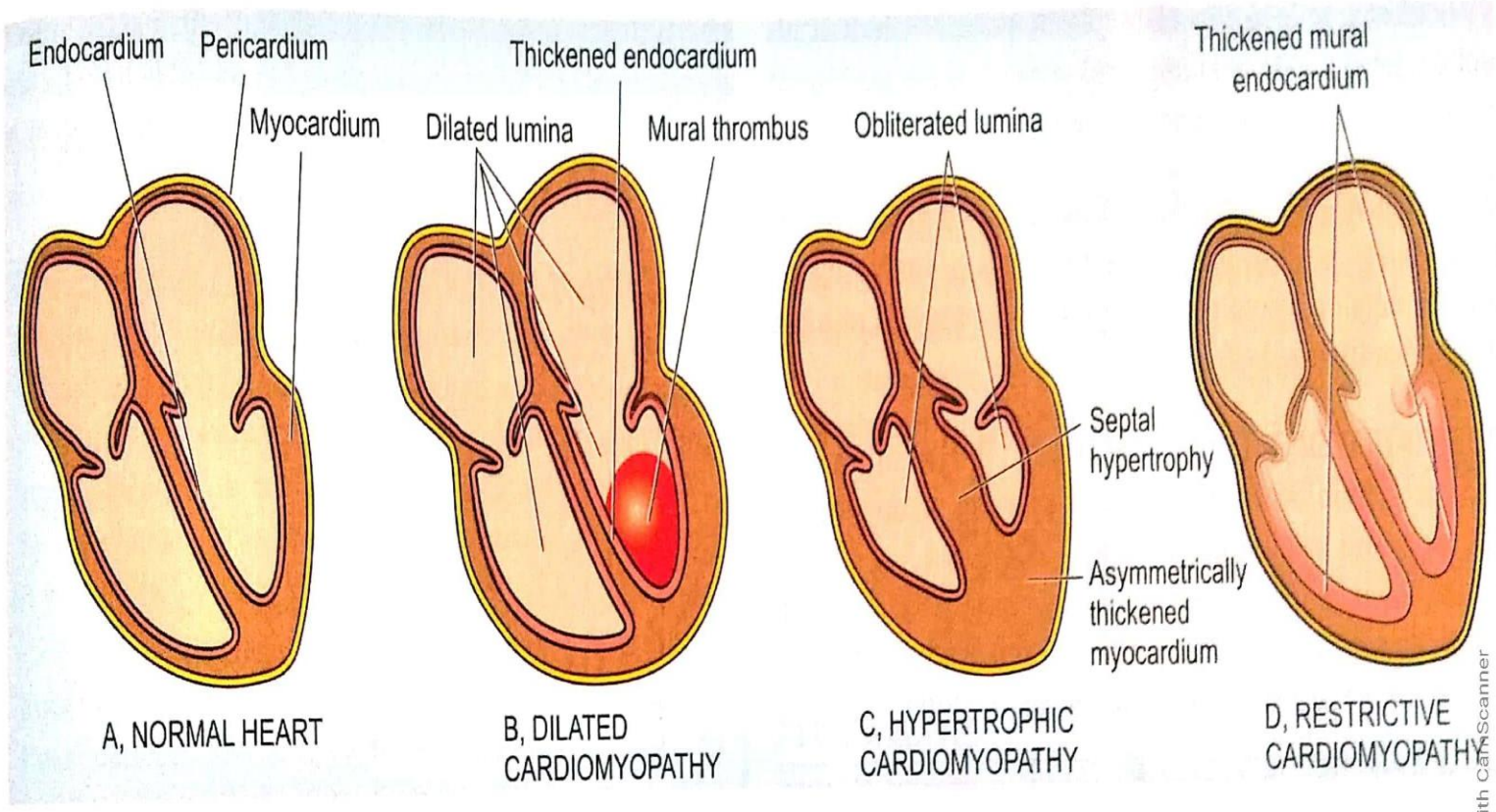


Figure 14.35 The major pathophysiologic forms of idiopathic cardiomyopathies.

This is referred to as 'alcoholic cardiomyopathy' and included ii) *Inherited mutations* in genes encoding for sarcome

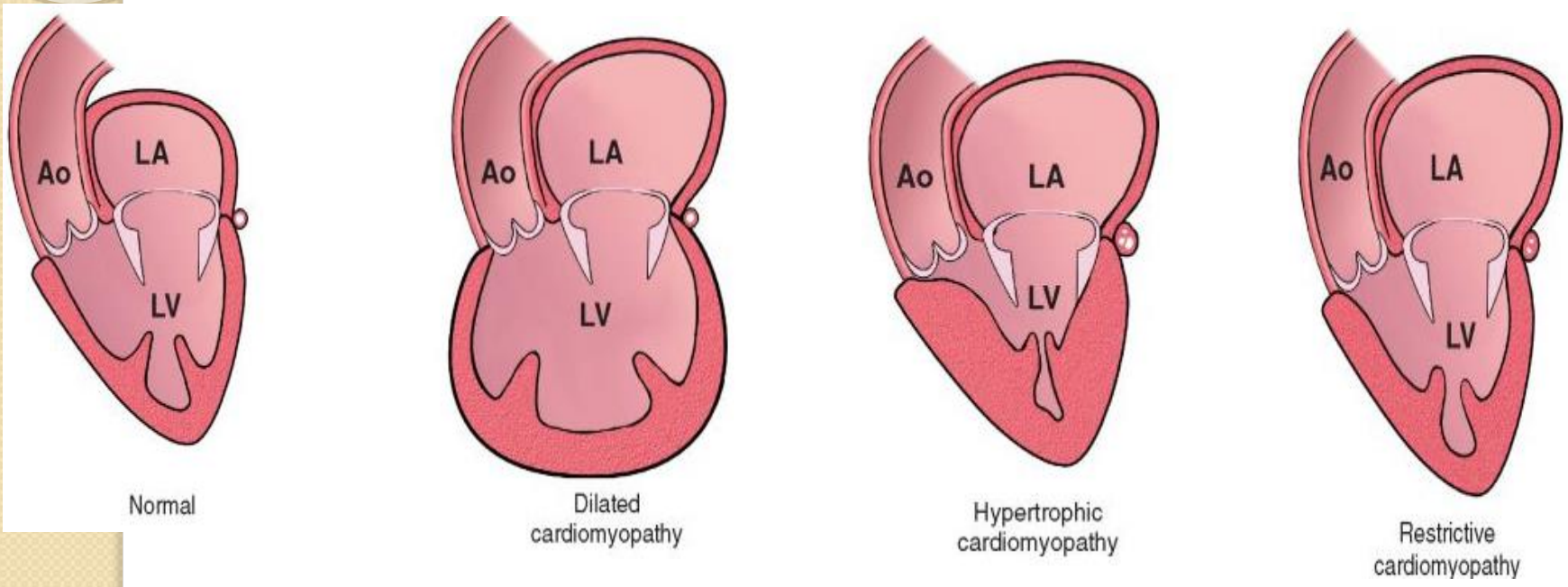
Table 14.10 Classification of primary cardiomyopathies.

1. IDIOPATHIC DILATED (OR CONGESTIVE) CARDIOMYOPATHY
2. IDIOPATHIC HYPERTROPHIC CARDIOMYOPATHY
   - i) Obstructive type
   - ii) Non-obstructive type
3. IDIOPATHIC RESTRICTIVE (OR OBLITERATIVE OR INFILTRATIVE) CARDIOMYOPATHY
   - i) Cardiac amyloidosis
   - ii) Endocardial fibroelastosis
   - iii) Endomyocardial fibrosis
   - iv) Löeffler's endocarditis (fibroplastic parietal endocarditis with peripheral blood eosinophilia)
   - v) Other forms

CARDIOMYOPATHY

- *Types of cardiomyopthies are determined by clinical, functional, and pathologic patterns present*
  - *Dilated cardiomyopathy (DCM)*
  - *Hypertrophic cardiomyopathy (HCM)*
  - *Restrictive cardiomyopathy*

CARDIOMYOPATHY



CARDIOMYOPATHY

Functional pattern	Left ventricular ejection fraction	Mechanism of heart failure	Causes of phenotype
Dilated	< 40%	**Impairment of contractility (systolic dysfunction)**	**Genetic; alcohol; peripartum; myocarditis; hemochromatosis; chronic anemia; doxorubicin (Adriamycin); sarcoidosis; idiopathic**
Hypertrophic	50 to 80%	**Impairment of compliance (diastolic dysfunction)**	**Genetic; Friedreich ataxia; storage diseases; infants of diabetic mother**
Restrictive	45 to 90%	**Impairment of compliance (diastolic dysfunction)**	**Amyloidosis; radiation-induced fibrosis; idiopathic**

CARDIOMYOPATHY

Conditions associated with cardiomyopathies are

- **Cardiac infections**
- **Toxins**
- **Metabolic**
- **Neuromuscular disease**
- **Storage disorders**
- **Infiltrative**
- **Immunological**

CARDIOMYOPATHY

Conditions associated with cardiomyopathies are

- ***Cardiac infections –***

  Viruses, Chlamydia, Rickettsia, Bacteria, Fungi, Protozoa

- ***Toxins***

  Alcohol, Cobalt, Catecholamines, Carbon monoxide , Lithium Hydrocarbons, Arsenic, Cyclophosphamide, Doxorubicin (Adriamycin) and daunorubicin

- ***Metabolic –***

  Hyperthroidism, Hypothyroidism, Hyperkalemia, Hypokalemia, Nutritional deficiency (protein, thiamine, other avitaminoses), Hemochromatosis

CARDIOMYOPATHY

Conditions associated with cardiomyopathies are

- ***Neuromuscular disorder –***

  Friedreich ataxia Muscular dystrophy Congenital atrophies

- ***Storage disorder and other depositions***

  Hunter-Hurler syndrome Glycogen storage disease Fabry disease Amyloidosis

- ***Infiltrative –***

  Leukemia Carcinomatosis Sarcoidosis Radiation-induced fibrosis

- ***Immunological –***

  Leukemia Carcinomatosis Sarcoidosis Radiation-induced fibrosis

CARDIAC INFECTIONS

- Viruses
- Chlamydia
- Rickettsia
- Bacteria
- Fungi
- Protozoa

TOXINS

- Alcohol
- Cobalt
- Catecholamines
- Carbon monoxide
- Lithium
- Hydrocarbons
- Arsenic
- Cyclophosphamide
- Doxorubicin (Adriamycin) and daunorubicin

METABOLIC

- Hyperthroidism
- Hypothyroidism
- Hyperkalemia
- Hypokalemia
- Nutritional deficiency (protein, thiamine, other avitaminoses)
- Hemochromatosis

NEUROMUSCULAR DISEASE

- Friedreich ataxia
- Muscular dystrophy
- Congenital atrophies

STORAGE DISORDERS AND OTHER DEPOSITIONS

- Hunter-Hurler syndrome
- Glycogen storage disease
- Fabry disease
- Amyloidosis

INFILTRATIVE

- Leukemia
- Carcinomatosis
- Sarcoidosis
- Radiation-induced fibrosis

IMMUNOLOGICAL

- Myocarditis (several forms)
- Post-transplant rejection

CARDIOMYOPATHY

- ***Endomyocardial biopsies** are used in the diagnosis and management of individuals with myocardial disease and in cardiac transplant recipients.*
- ***Endomyocardial biopsy involves inserting a device called a bioptome transvenously into the right side of the heart and using its jaws to snip a small piece of septal myocardium***

DILATED CARDIOMYOPATHY

- ***The term dilated cardiomyopathy (DCM) is applied to a form of cardiomyopathy characterized by progressive cardiac dilation and contractile (systolic) dysfunction, usually with concomitant hypertrophy.***

- ***It is sometimes called congestive cardiomyopathy.***

DILATED CARDIOMYOPATHY

Morphology

- ***Heart is usually enlarged, heavy (often weighing two to three times normal), and flabby, due to dilation of all chambers***
- ***Mural thrombi are common and may be a source of thromboemboli.***
- ***No primary valvular alterations, and mitral (or tricuspid) regurgitation, when present, results from left (or right) ventricular chamber dilation.***
- ***The histologic abnormalities in DCM are nonspecific and usually do not point to a specific etiologic agent.***

DILATED CARDIOMYOPATHY

PATHOGENESIS

- *DCM results due to many causes*

  - Genetic infleunces

  - Toxicities

  - Myocarditis- an inflammatory disorder that precedes the development of cardiomyopathy in at least some cases, and is sometimes caused by viral infections (Coxsackievirus B and other enteroviruses)

  - Postpartum

DILATED CARDIOMYOPATHY

PATHOGENESIS

Genetic causes –

- ***Autosomal-dominant inheritance** is the predominant pattern. X-linked, autosomal-recessive, and mitochondrial inheritance are less common*
- ***Deletions in mitochondrial genes that result in defects in oxidative phosphorylation***

DILATED CARDIOMYOPATHY

PATHOGENESIS

Genetic causes –

- ***X-linked cardiomyopathy is associated with mutations in the gene that encodes dystrophin***
- ***Other forms of DCM are associated with mutations in genes encoding cardiac α-actin (which links the sarcomere with dystrophin), desmin, and the nuclear lamina proteins, lamin A and lamin C.***

DILATED CARDIOMYOPATHY

PATHOGENESIS

Alcohol and other toxins

- ***Alcohol toxicity is due to direct ethanol toxicity or a secondary nutritional disturbance may be the cause of the myocardial injury.***
- ***Alcohol or its metabolites (especially acetaldehyde) have a direct toxic effect on the myocardium***
- ***chronic alcoholism may be associated with thiamine deficiency, which can lead to beriberi heart disease (also indistinguishable from DCM)***
- ***Doxorubicin (Adriamycin) and Cobalt causes myocardial insult***

DILATED CARDIOMYOPATHY

PATHOGENESIS

Peripartum cardiomyopathy

- ***occur late in pregnancy or several weeks to months postpartum***
- ***have been proposed as cPregnancy-associated hypertension, volume overload, nutritional deficiency, other metabolic derangements, or poorly characterized immunological reaction auses***
- ***Elevated levels of an anti-angiogenic cleavage product of the hormone prolactin (which rises late in pregnancy) and peripartum cardiomyopathy.***

DILATED CARDIOMYOPATHY

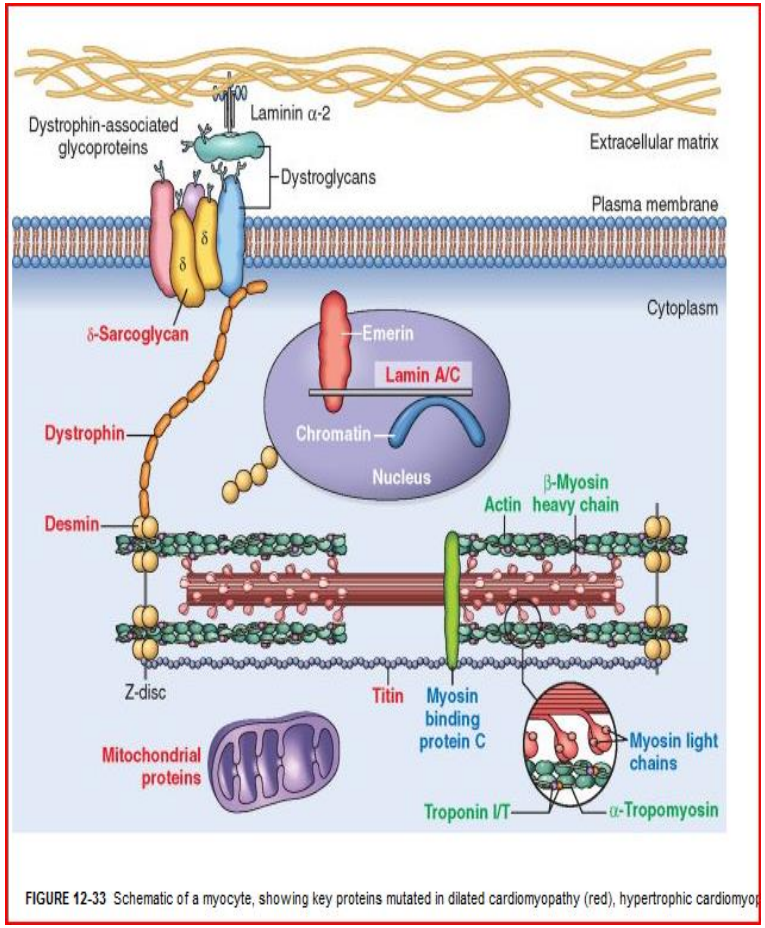
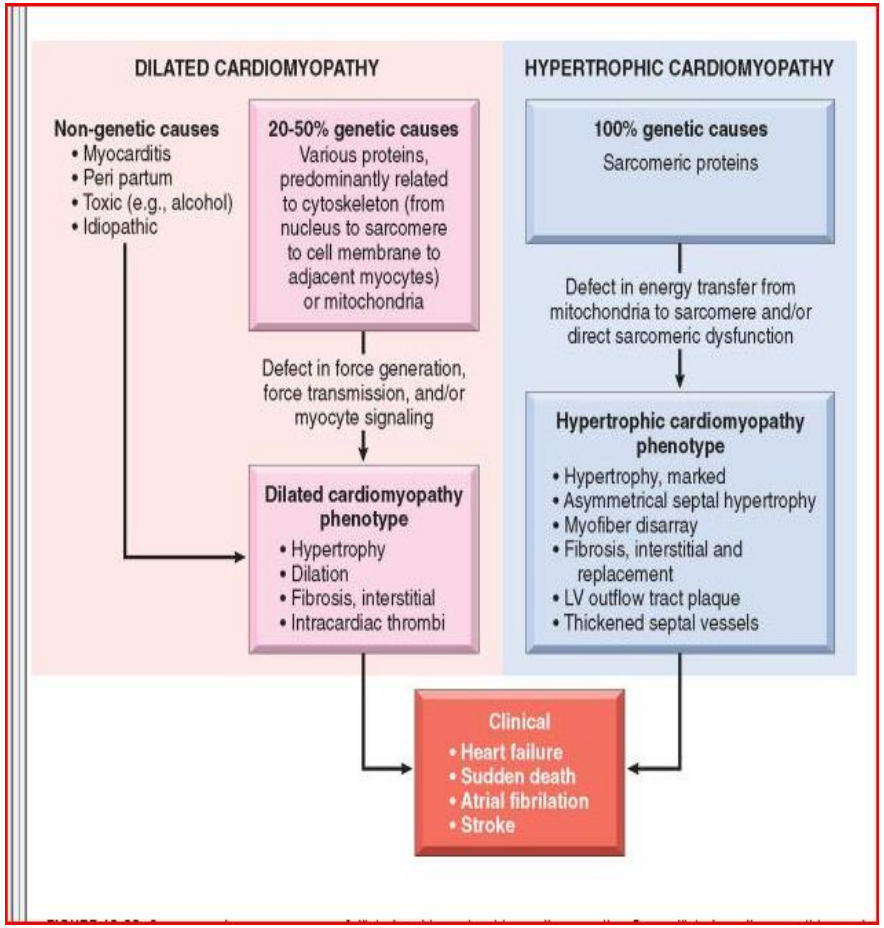


FIGURE 12-33 Schematic of a myocyte, showing key proteins mutated in dilated cardiomyopathy (red), hypertrophic cardiomyop

DILATED CARDIOMYOPATHY



DILATED CARDIOMYOPATHY

Clinical features

- ***most commonly affects individuals between the ages of 20 and 50.***
- ***Presents with slowly progressive signs and symptoms of CHF such as shortness of breath, easy fatigability, and poor exertional capacity.***
- ***In the end stage, patients often have ejection fractions of less than 25% (normal, ~50% to 65%).***

DILATED CARDIOMYOPATHY

Clinical features

- *Fifty percent of patients die within 2 years, and only 25% survive longer than 5 years*
- *Death is usually attributable to progressive cardiac failure or arrhythmia and can occur suddenly.*
- *Embolism from dislodgment of an intracardiac thrombus can occur*

HYPERTROPHIC CARDIOMYOPATHY

Hypertrophic cardiomyopathy (HCM) is characterized by

- *myocardial hypertrophy,*
- *poorly compliant left ventricular myocardium leading to abnormal diastolic filling,*
- *intermittent ventricular outflow obstruction (One third of cases)*

HCM is caused by mutations in genes encoding sarcomeric proteins

HYPERTROPHIC CARDIOMYOPATHY

Morphology

- ***The essential feature of HCM is massive myocardial hypertrophy, usually without ventricular dilation.***
- ***The classic pattern is disproportionate thickening of the ventricular septum as compared with the free wall of the left ventricle (with a ratio greater than 1 : 3), frequently termed asymmetric septal hypertrophy.***
- ***In about 10% of cases, however, the hypertrophy is symmetrical throughout the heart***

HYPERTROPHIC CARDIOMYOPATHY

Morphology

- ***Hypertrophy can involve the entire septum, it is usually most prominent in the subaortic region***
- ***On cross-section, the ventricular cavity loses its usual round-to-ovoid shape and may be compressed into a "banana-like" configuration by bulging of the ventricular septum into the lumen***

HYPERTROPHIC CARDIOMYOPATHY

Morphology

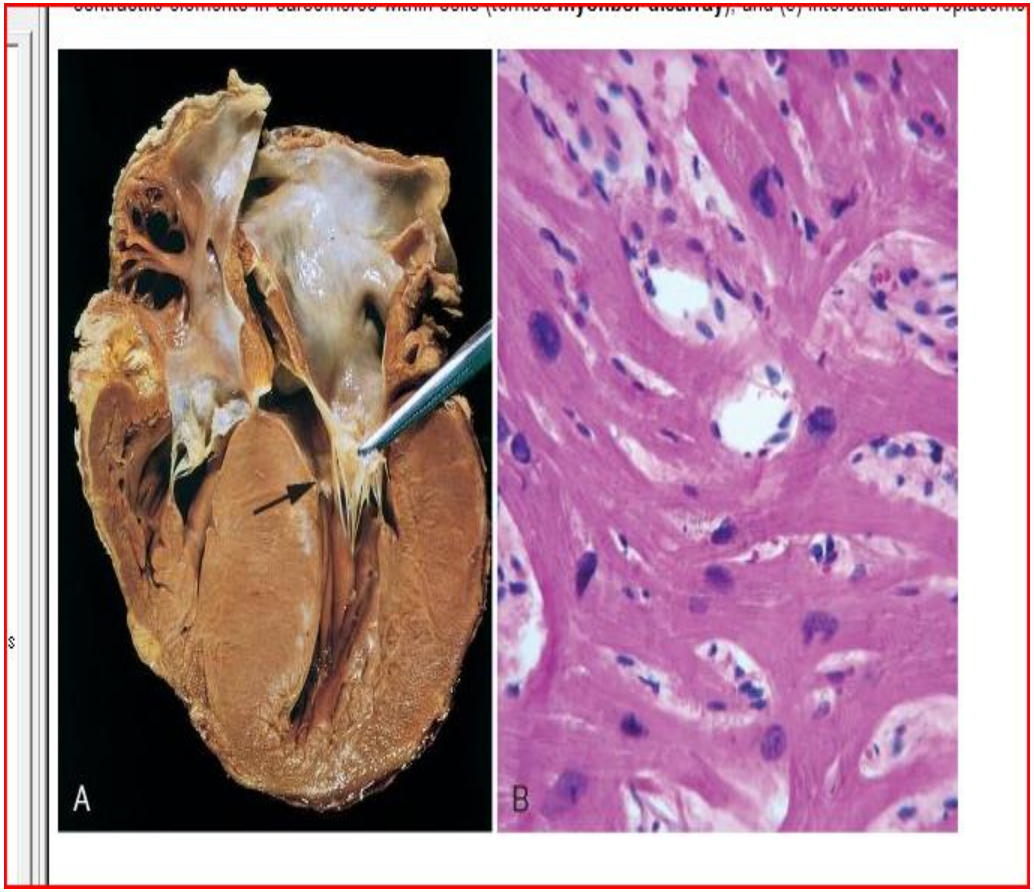
- ***Often present are endocardial thickening or mural plaque formation in the left ventricular outflow tract and thickening of the anterior mitral leaflet. Both findings are a result of contact of the anterior mitral leaflet with the septum during ventricular systole,***

HYPERTROPHIC CARDIOMYOPATHY

Histologic features of the myocardium in HCM are

- ***Extensive myocyte hypertrophy to a degree unusual in other conditions, with transverse myocyte diameters frequently greater than 40 μm (normal, ~15 μm)***
- ***Haphazard disarray of bundles of myocytes, individual myocytes, and contractile elements in sarcomeres within cells (termed myofiber disarray)***
- ***Interstitial and replacement fibrosis***

HYPERTROPHIC CARDIOMYOPATHY



HYPERTROPHIC CARDIOMYOPATHY

Pathogenesis

- ***caused by mutations in any one of several genes that encode sarcomeric proteins***
- ***Pattern of transmission is autosomal dominant***
- ***Mutations causing HCM are found most commonly in the gene encoding β-myosin heavy chain (β-MHC), cardiac TnT, α-tropomyosin, and myosin-binding protein C (MYBP-C)***

CARDIOMYOPATHY

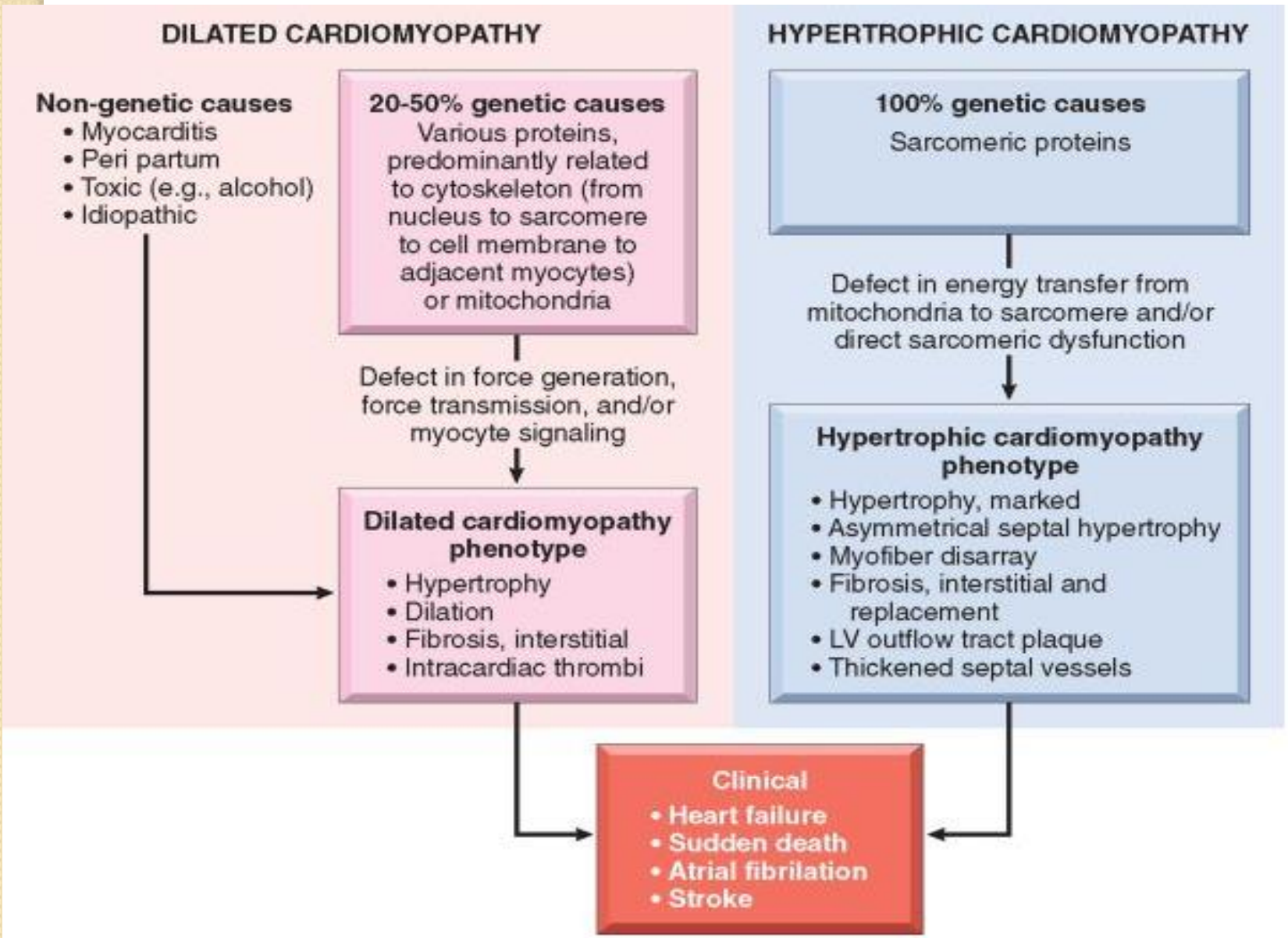
Clinical features

- ***The basic physiologic abnormality in HCM is reduced stroke volume due to impaired diastolic filling, which results from the reduced chamber size and compliance of the massively hypertrophied left ventricle***
- ***approximately 25% of patients have obstruction to the left ventricular outflow.***
- ***The limitation of cardiac output and a secondary increase in pulmonary venous pressure cause exertional dyspnea.***

HYPERTROPHIC CARDIOMYOPATHY

Clinical features

- ***The major clinical problems in HCM are atrial fibrillation, mural thrombus formation leading to embolization and possible stroke, intractable cardiac failure, ventricular arrhythmias, and, not infrequently, sudden death, especially in some affected families***
- ***HCM is one of the most common causes of sudden, otherwise unexplained death in young athletes***


DILATED CARDIOMYOPATHY
HYPERTROPHIC CARDIOMYOPATHY
Non-genetic causes
• Myocarditis
• Peri partum
• Toxic (e.g., alcohol)
• Idiopathic
20-50% genetic causes
Various proteins, predominantly related to cytoskeleton (from nucleus to sarcomere to cell membrane to adjacent myocytes) or mitochondria
100% genetic causes
Sarcomeric proteins
Defect in energy transfer from mitochondria to sarcomere and/or direct sarcomeric dysfunction
Defect in force generation, force transmission, and/or myocyte signaling
Dilated cardiomyopathy phenotype
• Hypertrophy
• Dilation
• Fibrosis, interstitial
• Intracardiac thrombi
Hypertrophic cardiomyopathy phenotype
• Hypertrophy, marked
• Asymmetrical septal hypertrophy
• Myofiber disarray
• Fibrosis, interstitial and replacement
• LV outflow tract plaque
• Thickened septal vessels
Clinical
• Heart failure
• Sudden death
• Atrial fibrilation
• Stroke

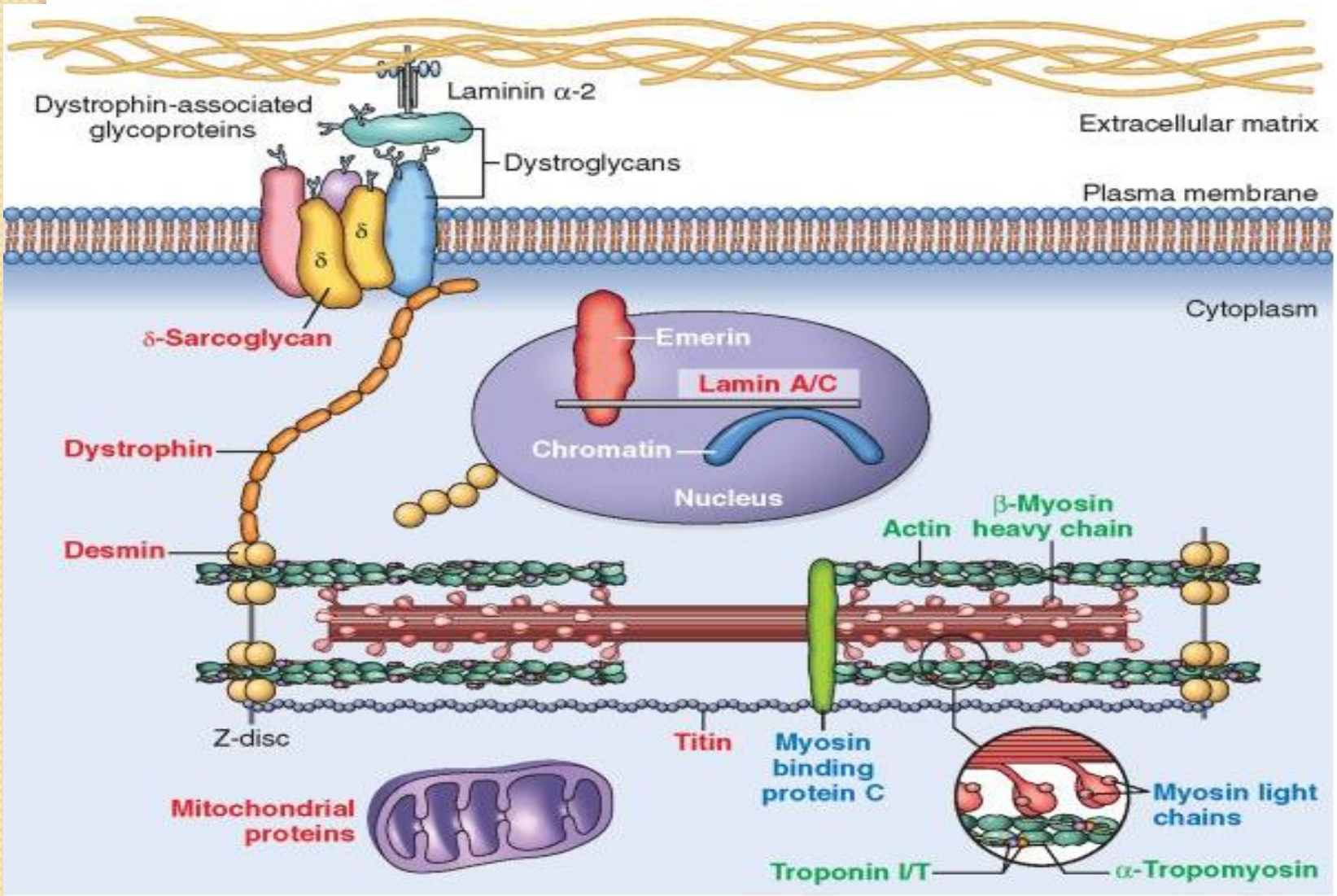


FIGURE 12-33 Schematic of a myocyte, showing key proteins mutated in dilated cardiomyopathy (red), hypertrophic cardiomyopathy (blue), or both (green).

RESTRICTIVE CARDIOMYOPATHY

- Primary decrease in ventricular compliance, resulting in impaired ventricular filling during diastole
- Systolic function of left ventricle unaffected

- Idiopathic
- Secondary to
  - Radiation fibrosis
  - Amyloidosis
  - Sarcoidosis
  - metastatic tumors
  - inborn errors of metabolism

- **Morphology :**
- Ventricles approximately normal size or slightly enlarged
- No dilatation
- Myocardium is firm and noncompliant
- Biatrial dilation is commonly observed

- **Microscopically :**
- Patchy or diffuse interstitial fibrosis, minimal to extensive
- Endomyocardial biopsy - specific etiology
- Associated Amyloidosis

Other restrictive conditions

- ***Endomyocardial fibrosis* :**
- Children
- Fibrosis of the ventricular endocardium and subendocardium - tricuspid and mitral valves
- Etiology is unknown
- ?? Organization of mural thrombi

- ***Loeffler endomyocarditis* :**

- Endomyocardial fibrosis, typically with large mural thrombi
- Peripheral eosinophilia
- Eosinophilic infiltrates in organs
- Major basic protein of eosinophils - endomyocardial necrosis
- Myeloproliferative disorder - PDGFRα or PDGFRβ gene rearrangements

- ***Endocardial fibroelastosis :***

- Obscure etiology
- Focal or diffuse fibroelastic thickening
- *M/C* - mural left ventricular endocardium
- *M/C in first 2 years of life – assoc. congenital cardiac disease*

MYOCARDITIS

- Primary – Infectious etiology
- Secondary – IHD

- *Viral infections* are the most common cause
- *Coxsackieviruses A* and *B* and other enteroviruses
- ?? Direct injury / destructive immune response

TABLE 12-12 -- Major Causes of Myocarditis

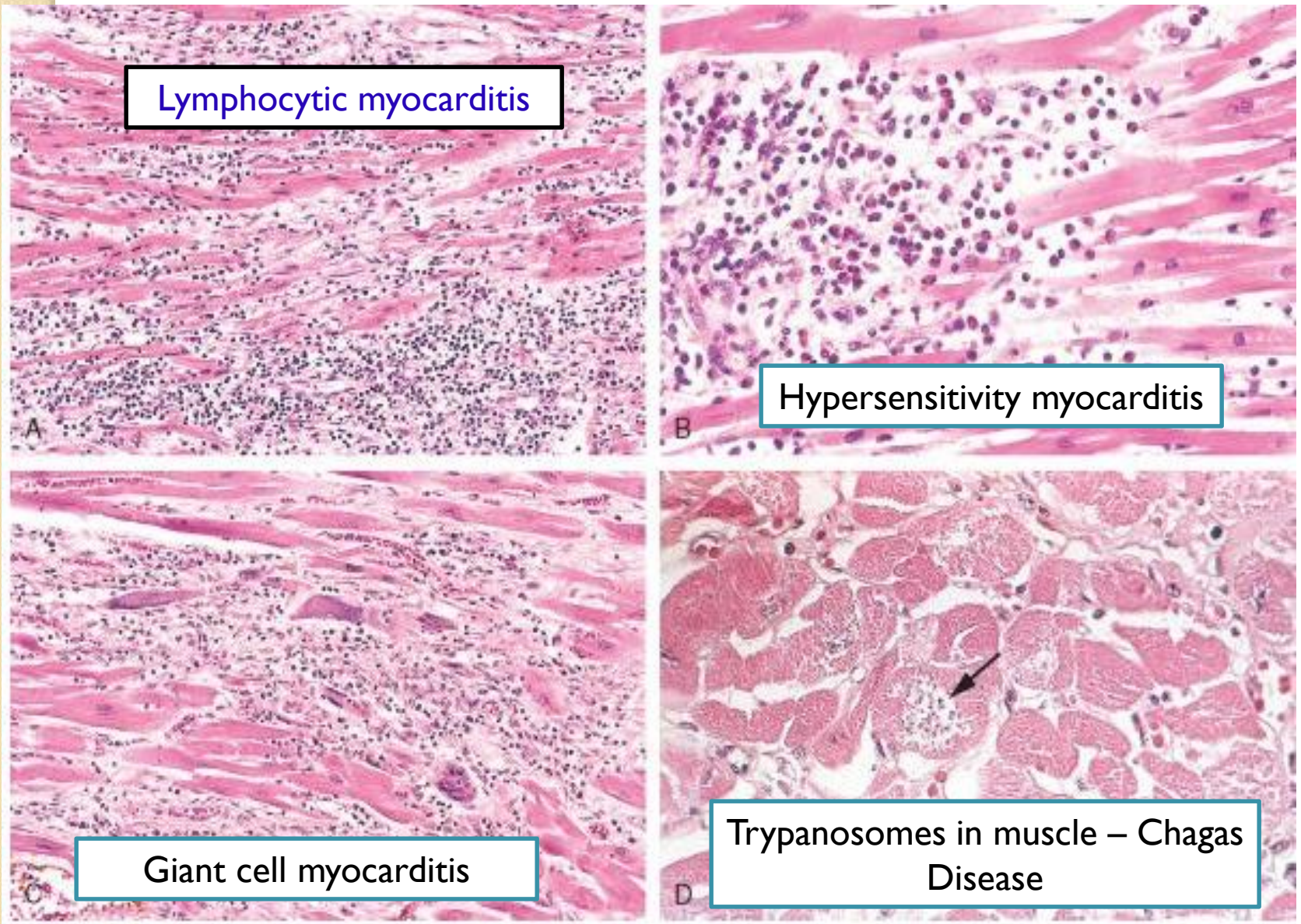
INFECTIONS
Viruses (e.g., coxsackievirus, ECHO, influenza, HIV, cytomegalovirus)
Chlamydiae (e.g., *C. psittaci*)
Rickettsiae (e.g., *R. typhi*, typhus fever)
Bacteria (e.g., *Corynebacterium diphtheriae, Neisseria meningococcus, Borrelia* (Lyme disease)
Fungi (e.g., *Candida*)
Protozoa (e.g., *Trypanosoma cruzi* [Chagas disease], toxoplasmosis)
Helminths (e.g. trichinosis)
IMMUNE-MEDIATED REACTIONS
Postviral
Poststreptococcal (rheumatic fever)
Systemic lupus erythematosus
Drug hypersensitivity (e.g., methyldopa, sulfonamides)
Transplant rejection
UNKNOWN
Sarcoidosis
Giant cell myocarditis

HIV, human immunodeficiency virus.

Morphology

- Active phase – Heart normal or dilated
- Advanced stages – Myocardium flabby
- Hemorrhagic lesions
- Mural thrombi

- **Microscopy :**
- Interstitial mononuclear inflammatory infiltrate
- Focal or diffuse
- Focal myocyte necrosis - fibrosis


Lymphocytic myocarditis
A
B
Hypersensitivity myocarditis
C
Giant cell myocarditis
D
Trypanosomes in muscle – Chagas Disease

Clinical Features

- Asymptomatic
- Precipitous onset of heart failure or arrhythmias – sudden death

Pericardial Disease

- Normally, there are about 30 to 50 mL of thin, clear, straw-colored fluid in the pericardial sac

- **Distension of parietal pericardium :**
- Serous fluid *(pericardial effusion)*
- Blood (hemopericardium), or
- Pus (purulent pericarditis)

PERICARDITIS

TABLE 12-13 -- Causes of Pericarditis

INFECTIOUS AGENTS
Viruses
Pyogenic becteria
Tuberculosis
Fungi
Other parasites
PRESUMABLY IMMUNOLOGICALLY MEDIATED
Rheumatic fever
Systemic lupus erythematosus
Scleroderma
Postcardiotomy
Postmyocardial infarction (Dressler) syndrome
Drug hypersensitivity reaction
MISCELLANEOUS
Myocardial infarction
Uremia
Following cardiac surgery
Neoplasia
Trauma
Radiation

- **Fibrinous and Serofibrinous Pericarditis :**
- Serous fluid mixed with a fibrinous exudate
- Common causes include acute MI, the postinfarction (Dressler) syndrome,uraemia, atoimmune causes
- Deposition of leucocytes and RBC
- Exudative fluid

- **Purulent or Suppurative Pericarditis :**
- Invasion of the pericardial space by microbes

  (1) Direct extension from neighboring infections, such as an empyema of the pleural cavity, lobar pneumonia, mediastinal infections, or extension of a ring abscess through the myocardium or aortic root

  (2) Seeding from the blood

  (3) Lymphatic extension; or

  (4) Direct introduction during cardiotomy

- Immunosuppression predisposes to infection

- Acute inflammatory reaction
- *Mediastinopericarditis*
- *Constrictive pericarditis as a consequence*

- **Hemorrhagic Pericarditis :**
- Exudate is composed of blood mixed with a fibrinous or suppurative effusion
- M/C Malignancy
- Other – bacterial infections, TB, Bleeding diathesis

- **Caseous Pericarditis :**
- TB
- Fungal infections
- Constrictive pericarditis - consequence

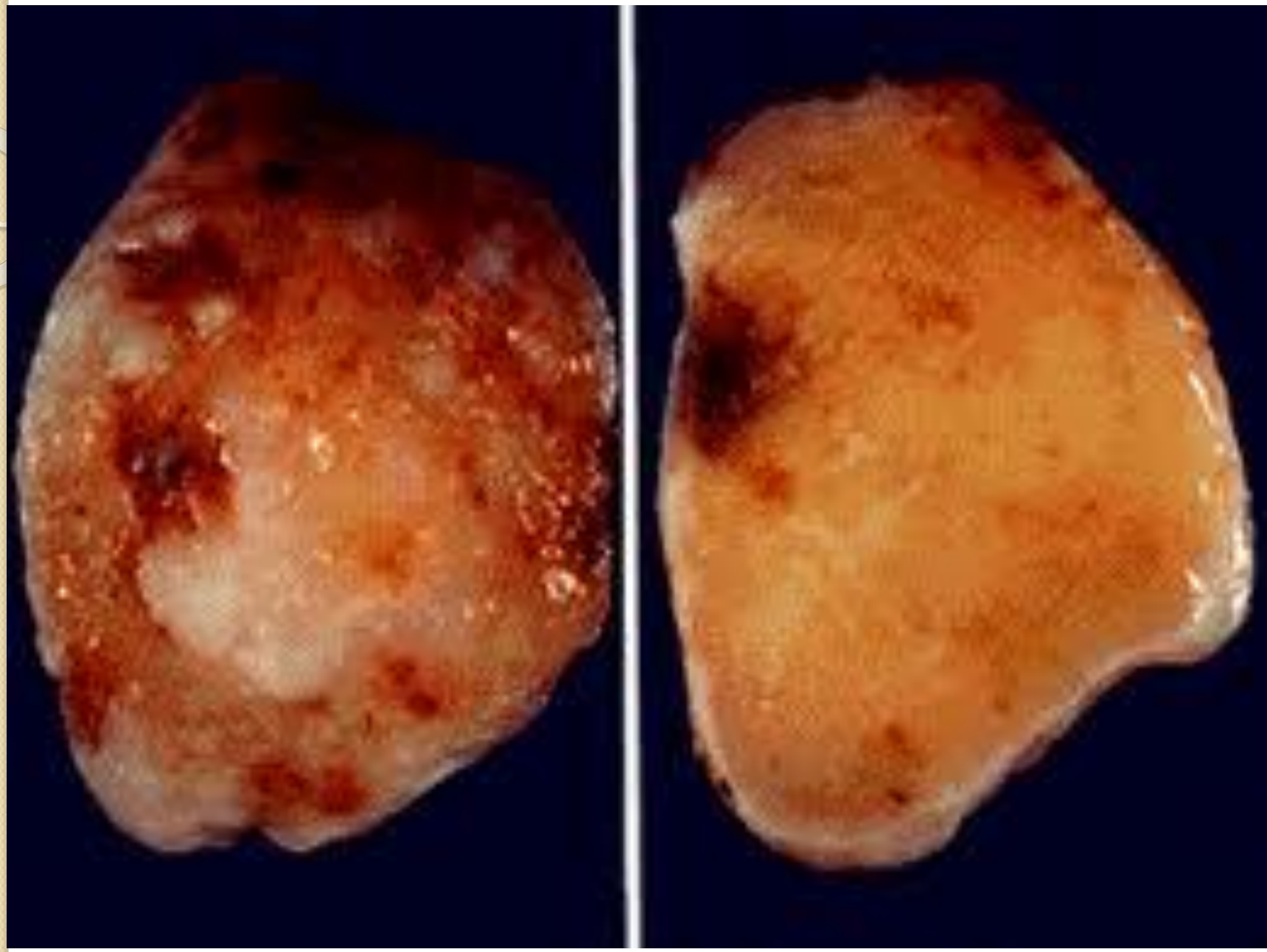
- **Chronic or Healed Pericarditis :**
- Organization - plaquelike fibrous thickenings - “soldier's plaque”
- Thin stringy adhesions
- *Adhesive mediastinopericarditis*
- *Constrictive pericarditis* - the heart is encased in a dense, fibrous or fibrocalcific scar that limits diastolic expansion and cardiac output

Myxoma

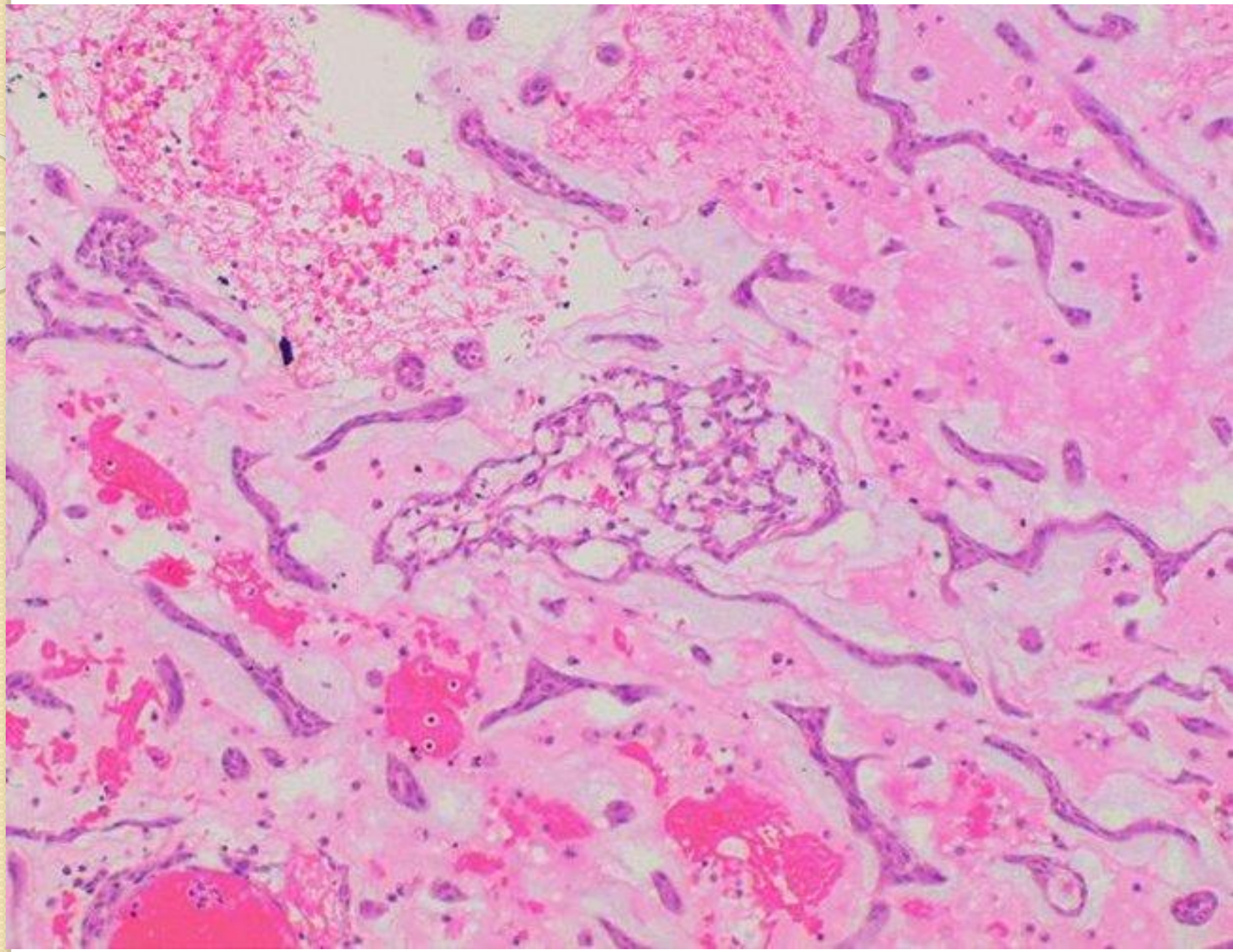
- Most common primary tumor of the heart
- Benign
- Associated with clonal abnormalities of Ch 12 & 17
- Any chamber. 90% atria
- Left: right – 4:1

Morphology

- Usually single
- Fosssa ovalis
- 1-10 cms
- Sessile or pedunculated
- Globular hard masses mottled with hemorrhage to soft, translucent, papillary or villious lesions with gelatinous appearance



- Stellate cells embedded within an abundant mucopolysaccharide ground substance
- Peculiar vessel like or gland like structures are characteristic
- Hemorrhages and mononuclear cells are commonly present



Clinical features

- Valvular ball-valve obstruction, embolization or fever & malise
- Echocardiography
- Surgery

Carney Complex

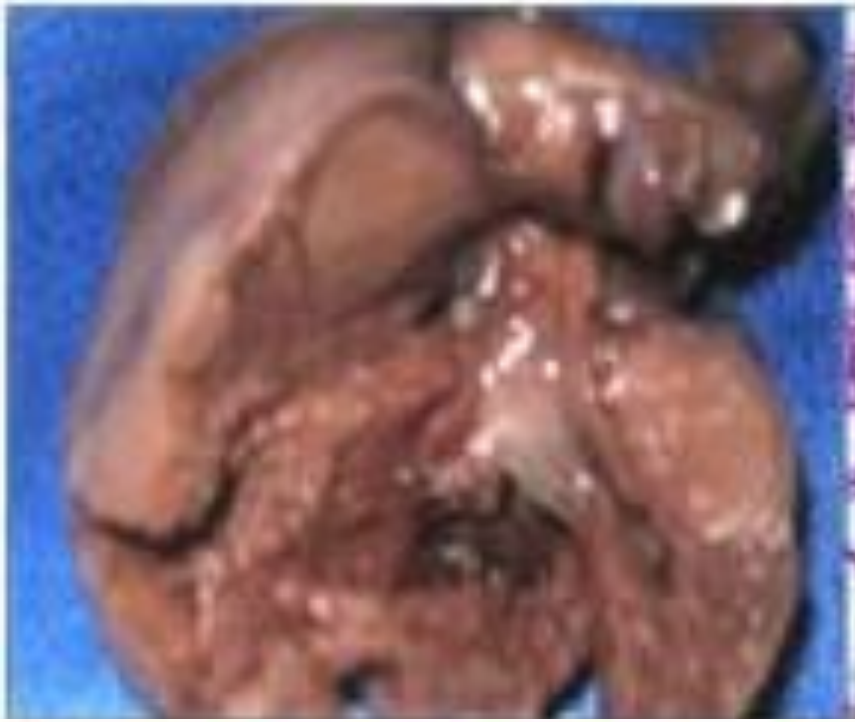
- AD
- Multiple cardiac & often extracardiac (skin) myxomas, pigmented skin lesions and endocrine overactivity

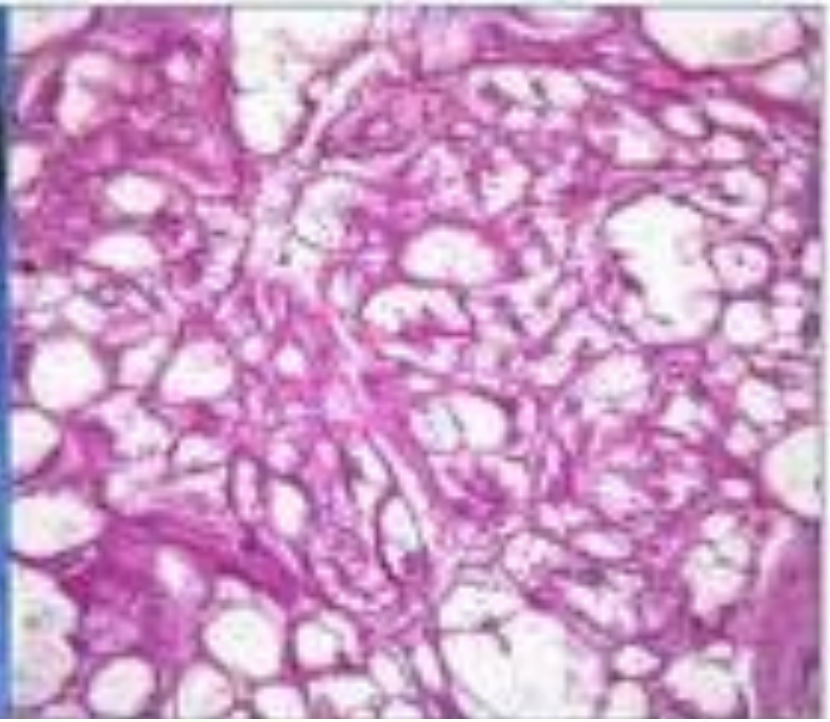
- PRKAR1 on Ch 17 – encode C-AMP dependent protein kinase A is mutated

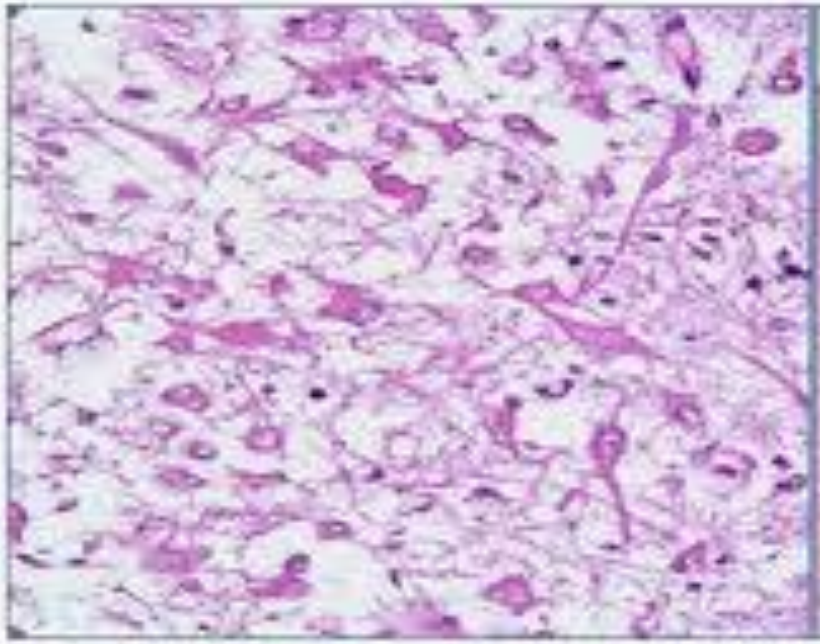
Rhabdomyoma

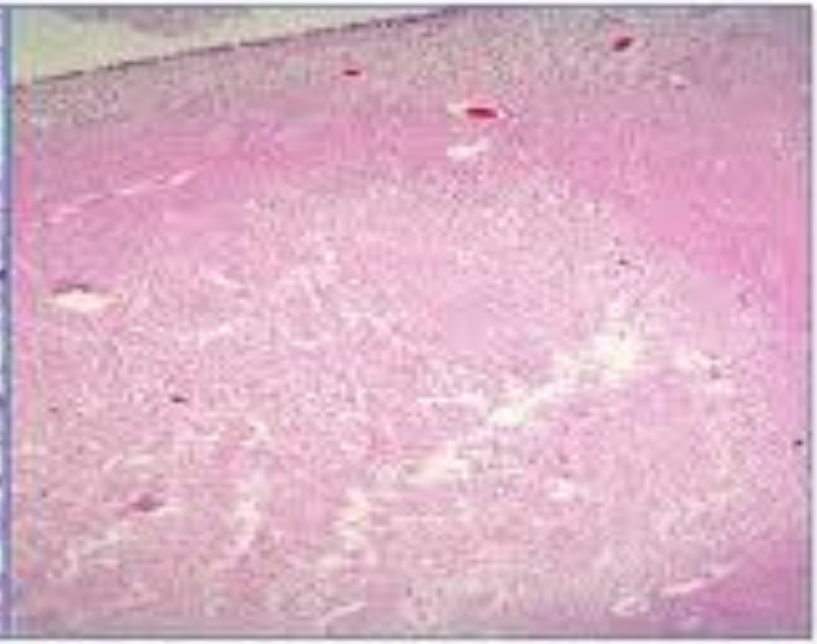
- Most frequent primary tumor of heart in infants & children
- Often associated with Tuberuos Sclerosis
- Defects in TSC1 & TSC2 genes

- Small gray-white myocardial masses
- Usually multiple in number, involves ventricles mainly protruding into the chamber
- Bizarre markedly enlarged myocytes
- Cytoplasm is reduced to thin webs or strands that extend to cell membranes – spider cells



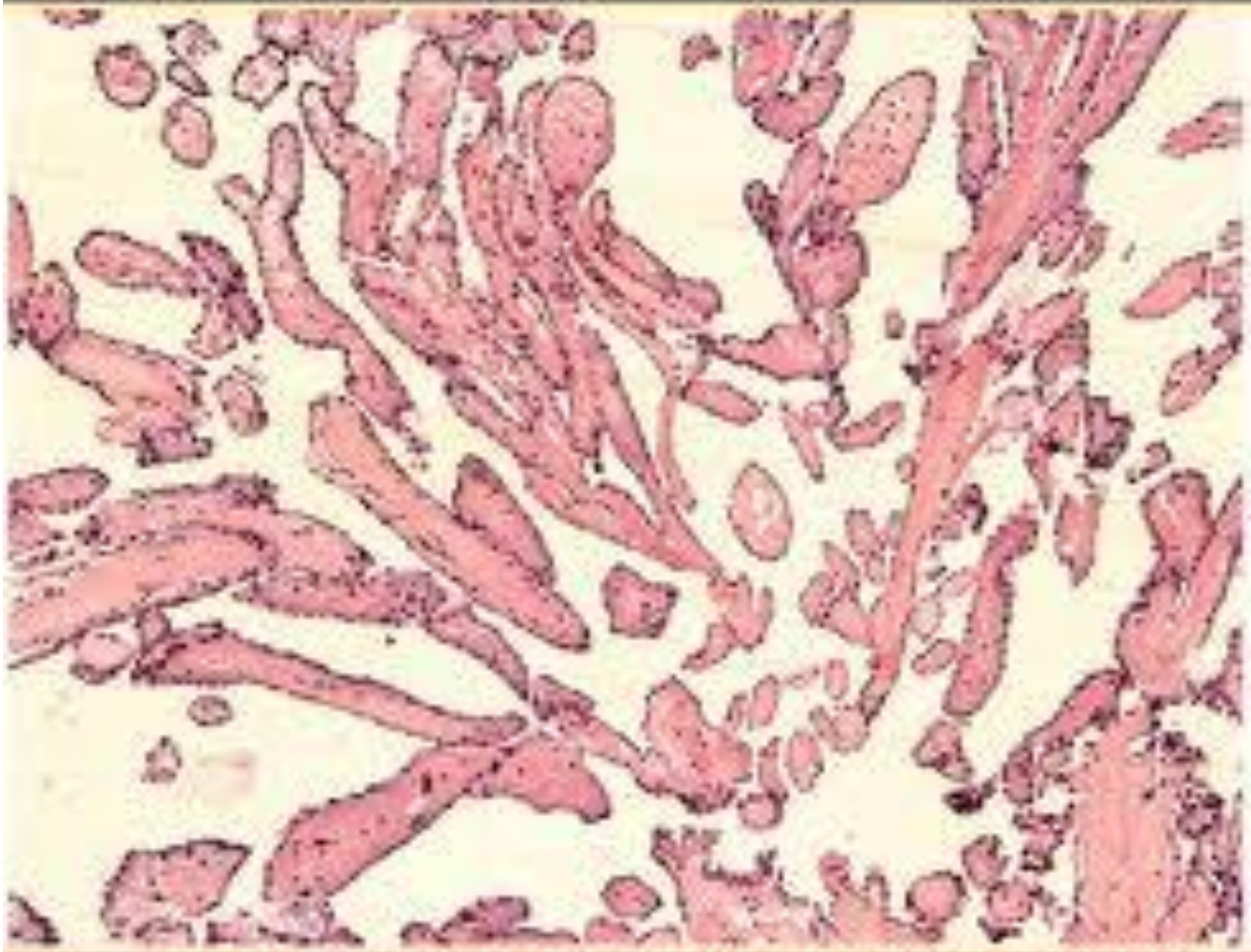






Papillary fibroelastoma

- Incidental seanemone like lesions
- May embolize
- Unusual benign neoplasms
- Located on valves on ventricular surface
- Cluster of hair like projections upto 1cm in length
- Core of myxoid tissue containing abundant mucopolysaccharides and elastic fibers covered by endothelium



Hemangioma

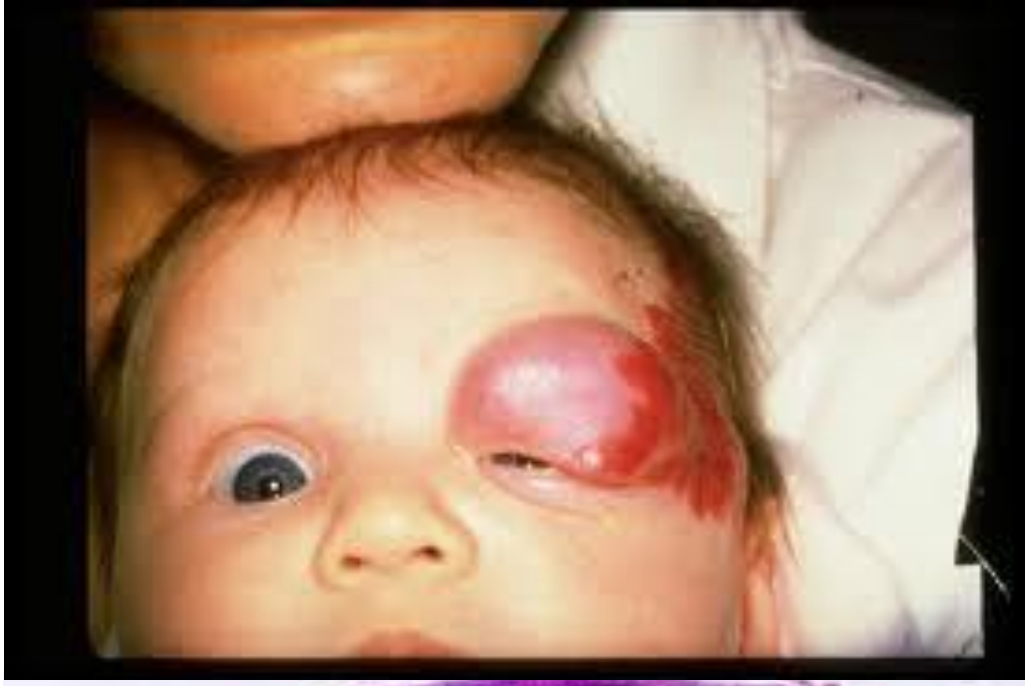
- Very common tumors
- Increased number of normal or abnormal vessels filled with blood
- 7% of childhood neoplasms
- Regress spontaneously

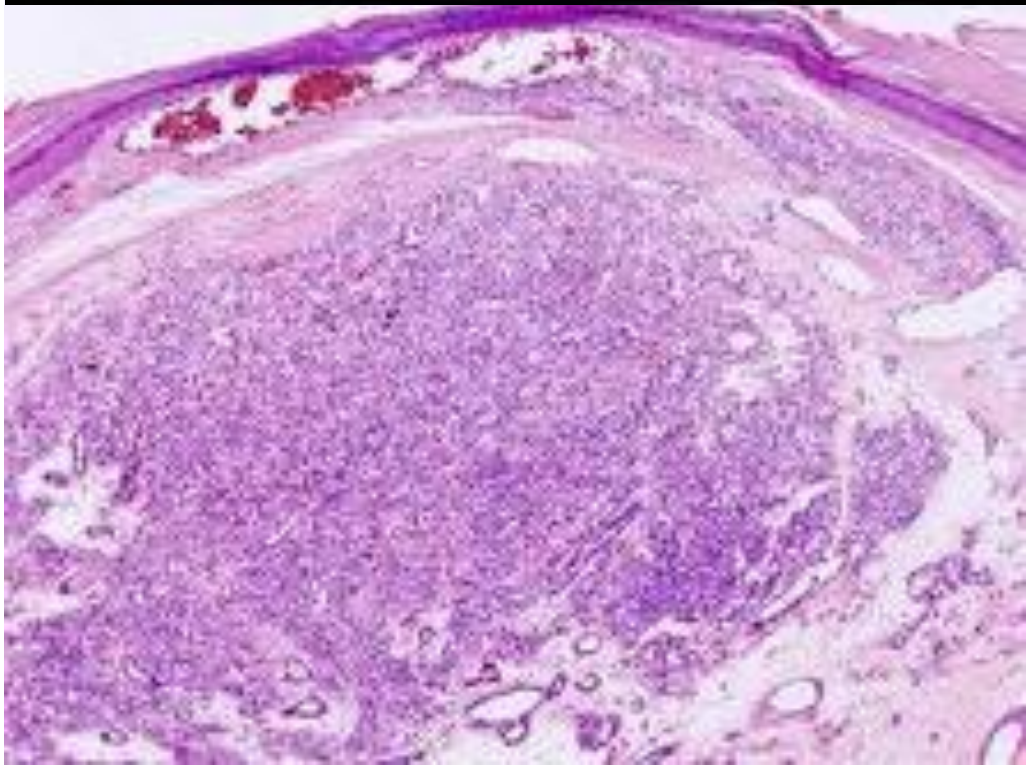
Angiomatosis:

- Involves large portions of the body
- Majority superficial, often heads & neck
- Liver

Capillary hemangiomas

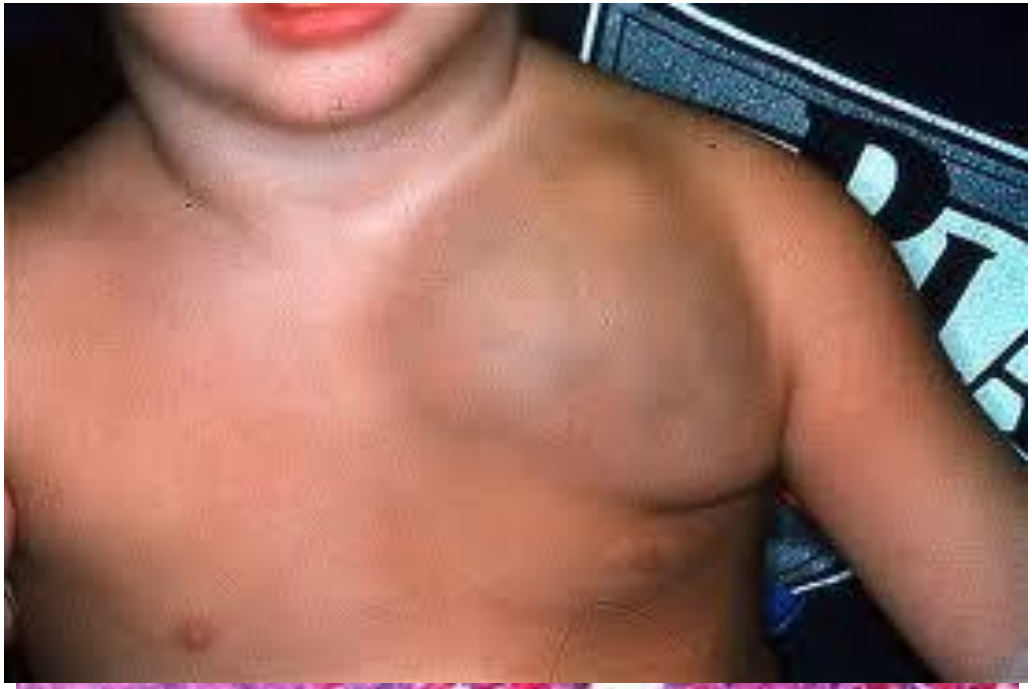
- Most common variant
- Skin, subcutaneous tissues, mucus membranes of oral cavities & lips
- Liver, spleen & kidney
- Unencapsulated aggregates of closely packed thin walled capillaries, blood filled & lined by endothelium
- Scant connective tissue

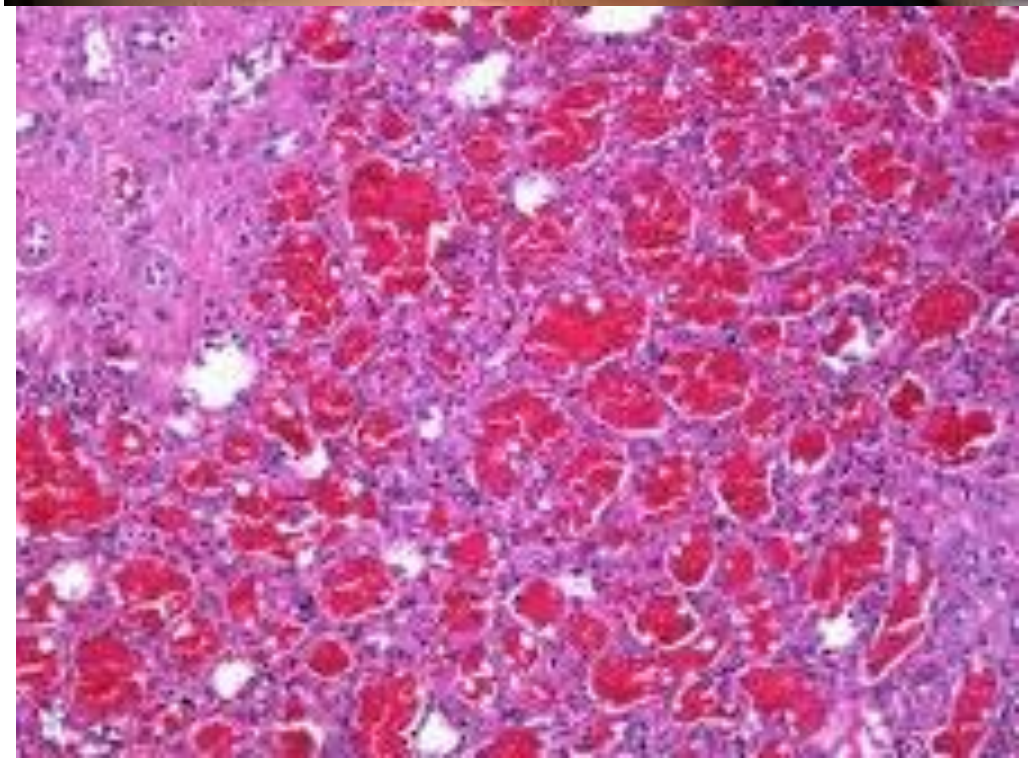




Cavernous hemangiomas

- Large dilated vascular channels
- Less well circumscribed
- More frequently located in deep structures
- No spontaneous regression
- Vulnerable to traumatic ulceration, cosmetic disturbances
- Brain hemangiomas – more dangerous



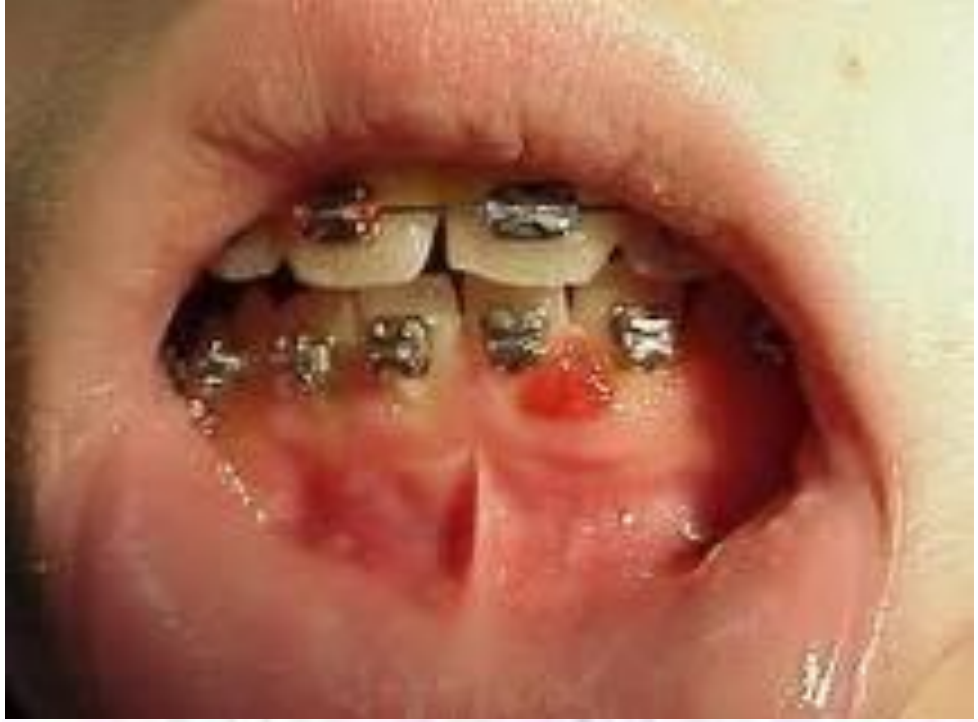


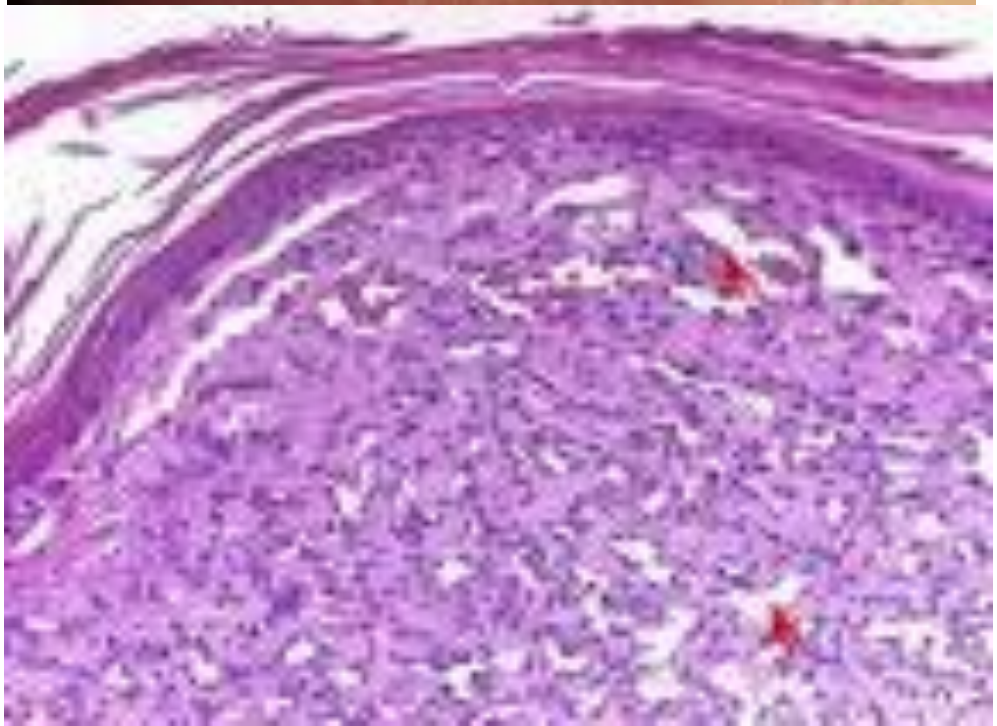
VHL disease –

- Cerebellum, brainstem or retina
- Cystic neoplasms in pancreas & liver
- Renal neoplasms

Pyogenic granulomas

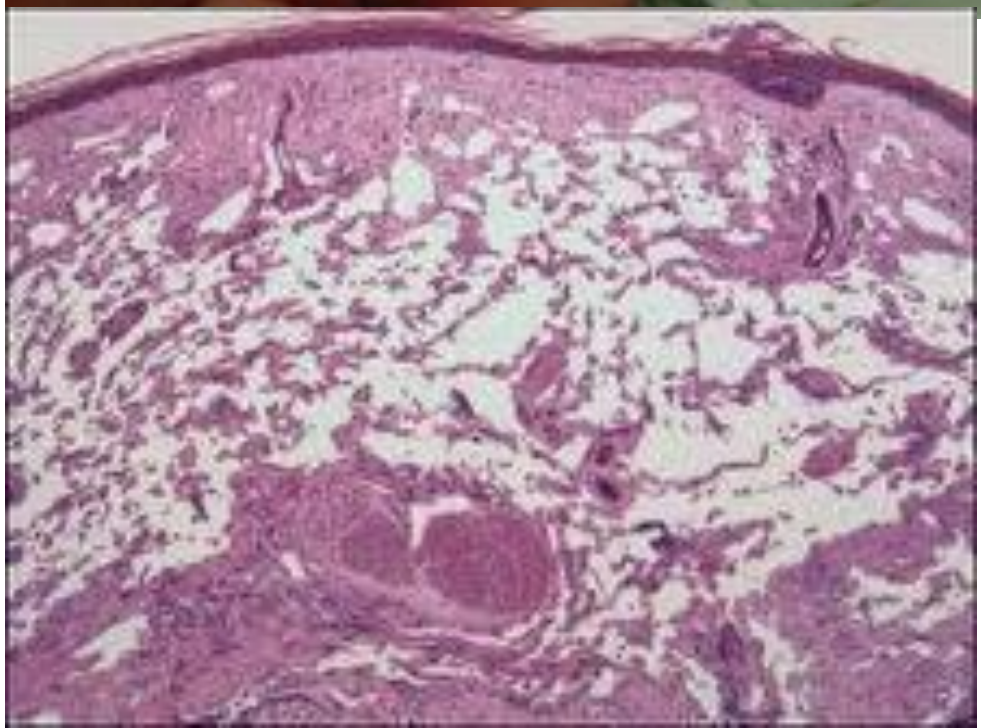
- Form of capillary hemangiomas
- Rapidly growing pedunculated red nodule on the skin or gingiva or oral mucosa
- 1/3rd develop after trauma
- Proliferating capillaries, edema & acute & chronic inflammatory cells
- Pregnancy tumor (granuloma gravidarum)
- Gingiva of pregnant women
- Spontaneously regress or undergo fibrosis





Lymphangioma

- Capillary lymphangioma
- Cavernous lymphagioma (cystic hygroma)

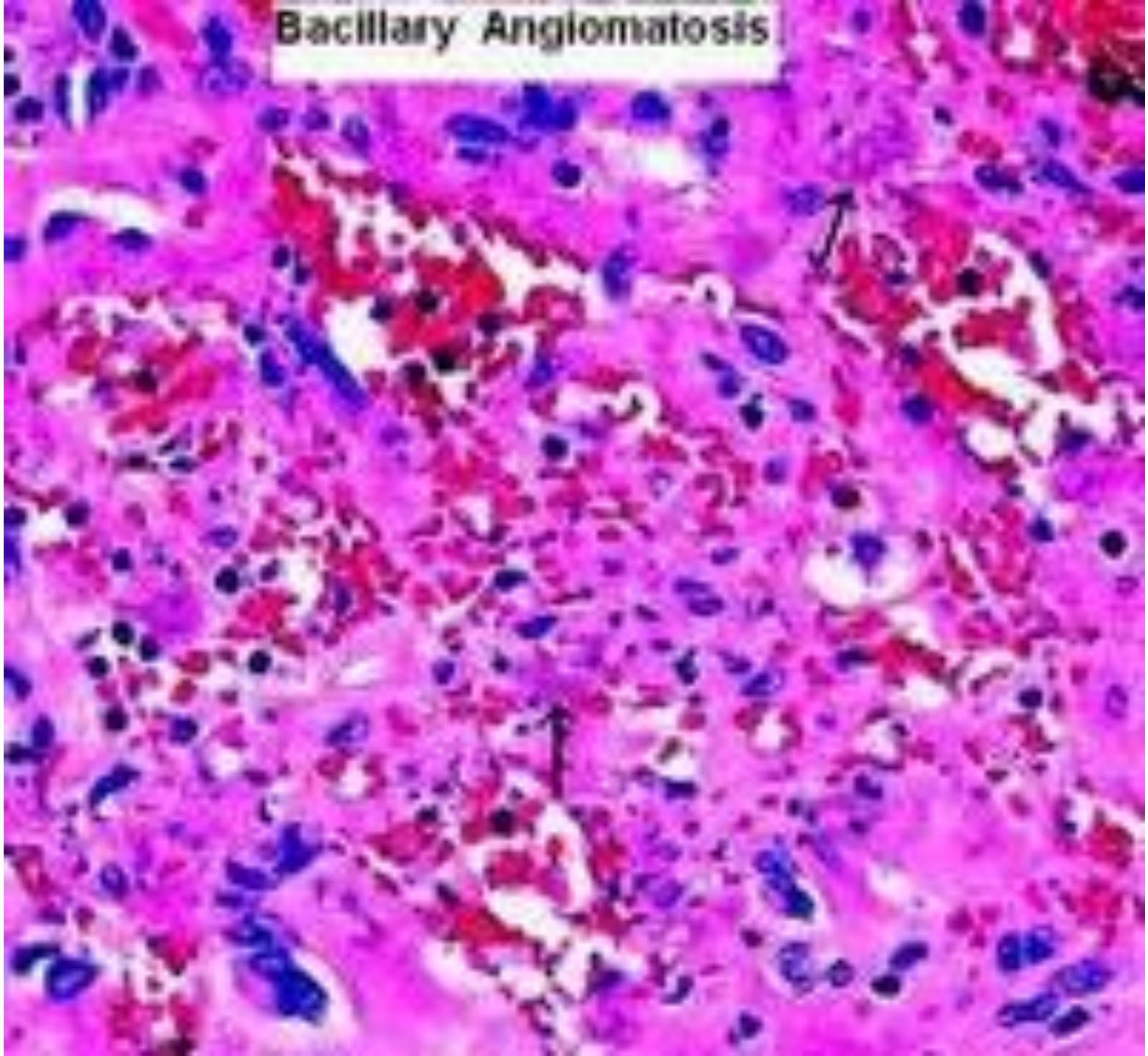


Bacillary angiomatosis

- First described in AIDS patients
- An opportunistic infection manifest as vascular proliferations resembling tumor
- Skin, bone, brain & other organs
- Gram negative organisms Bartonella family (B.Hensle, B.quintana)

Morphology

- One to numerous red papules & nodules
- Proliferation of capillaries that exhibit protuberant epitheliod endothelial cells, with nuclear atypia and mitosis
- Numerous stromal neutrophils, nuclear dust and purplish granular material consisting of the causative bacteria

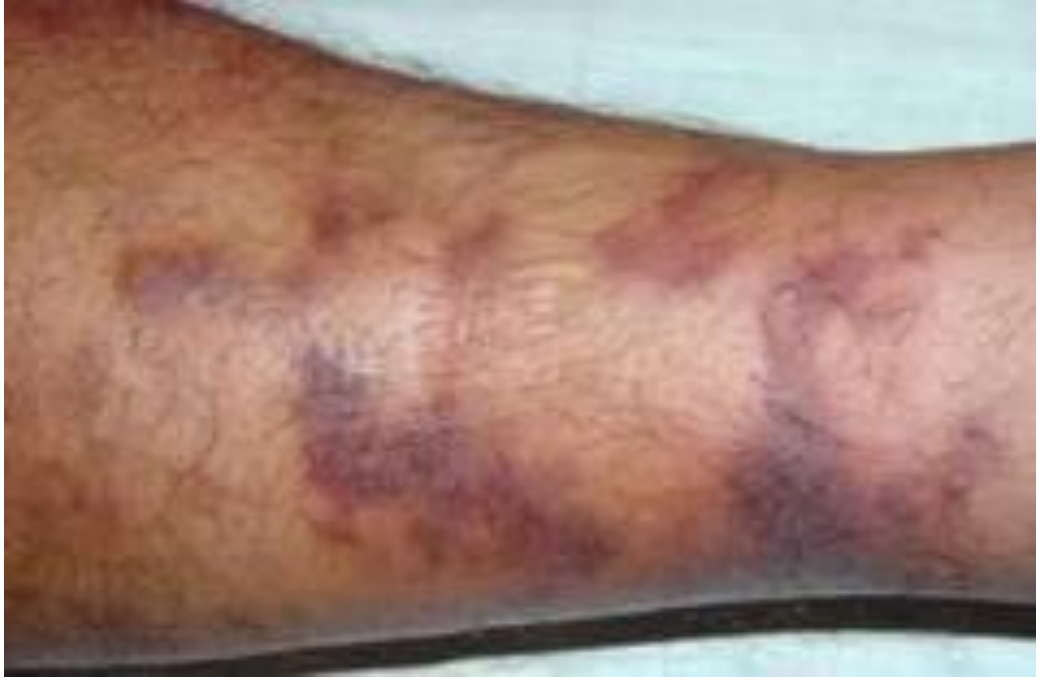

Bacillary Angiomatosis

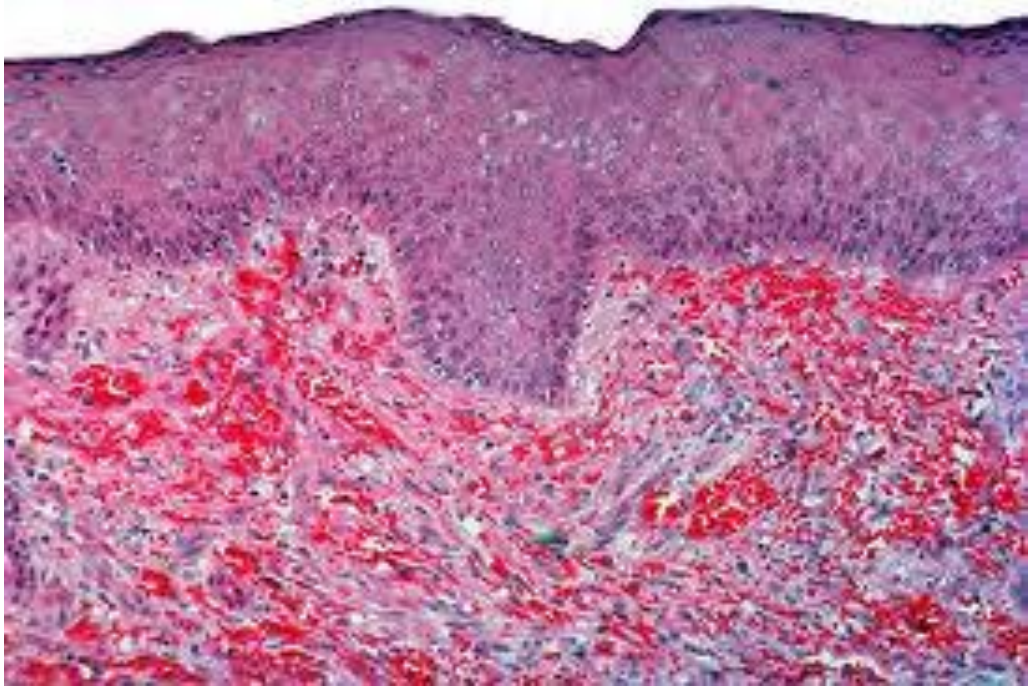
Kaposi sarcoma

- Frequently occurs in AIDS patients
- Fours forms of the disease are recognized

Chronic/classic or European KS

- 1872
- Older men
- Not associated with HIV
- Multiple red to purple skin plaques or nodules, primarily on arms and legs slowly increase in number & size
- Asymptomatic, localized to skin & subcutaneous tissue





Lymphadenopathic/African/Endemic KS

- Young, Bantu children os south Africa
- Localized, or generalized lymphadenopathy
- Extremely aggressive
- Skin lesions are sparse

Transplant-associated (immunosupression associated KS

- Solid organ transplantation
- Aggressive
- Involves lymphnodes, mucosa & visceral organs
- Skin lesions may be absent
- Sometimes regress

AIDS associated KS

- US
- 1/3rd of AIDS patient
- Now reduced in HAART Rx
- No site predilection, but involvement of lymphnodes & gut and wide dissemination is common early in the disease
-

Morphology

- In classic disease three stages can be identified
- Patch, plaque & nodule

Patch

- Pink to red to purple solitary or multiple macules, usually confined to distal extremities or feet
- Dilated, irregular & angulated blood vessels lined by endothelial cells, interspersed with lymphocytes, plasma cells & macrophages

Plaques

- Large, violacious & raised
- Dermal, dilated, jagged vascular channels lined by plump spindle cells with perivascular aggregates of spindle cells
- Scattered between channels are lymphocytes, hemosiderin laden macrophages
- pink hyaline globules are seen

Nodules

- More distinctly neoplastic
- Sheets of plump proliferating spindle cells mostly in the dermis
- Scattered small vessels & slit like spaces containing red cells are seen
- Hemorrhage, hemosiderin, lymphocytes
- Mitosis common
- Pink cytoplasmic globules
- Often accompanied by involvement of lymphnodes & viscera

Pathogenesis

- 1994
- HHV-8 virus/KSHV
- 95%
- Necessary and sufficient for the development of the disease
- Immunosupression appears to be an important cofactor
- KHSV is sexually transmitted

Clinical features

- Mostly assymptomatic
- Classic form is restricted to surface of the body
- Resection
- Radiation for multiple lesions
- Endemic KS is treated with chemotherapy
- In immunsuoression associated cases withdrawal of the immunosupression
- In AIDS associated cases retroviral Rx

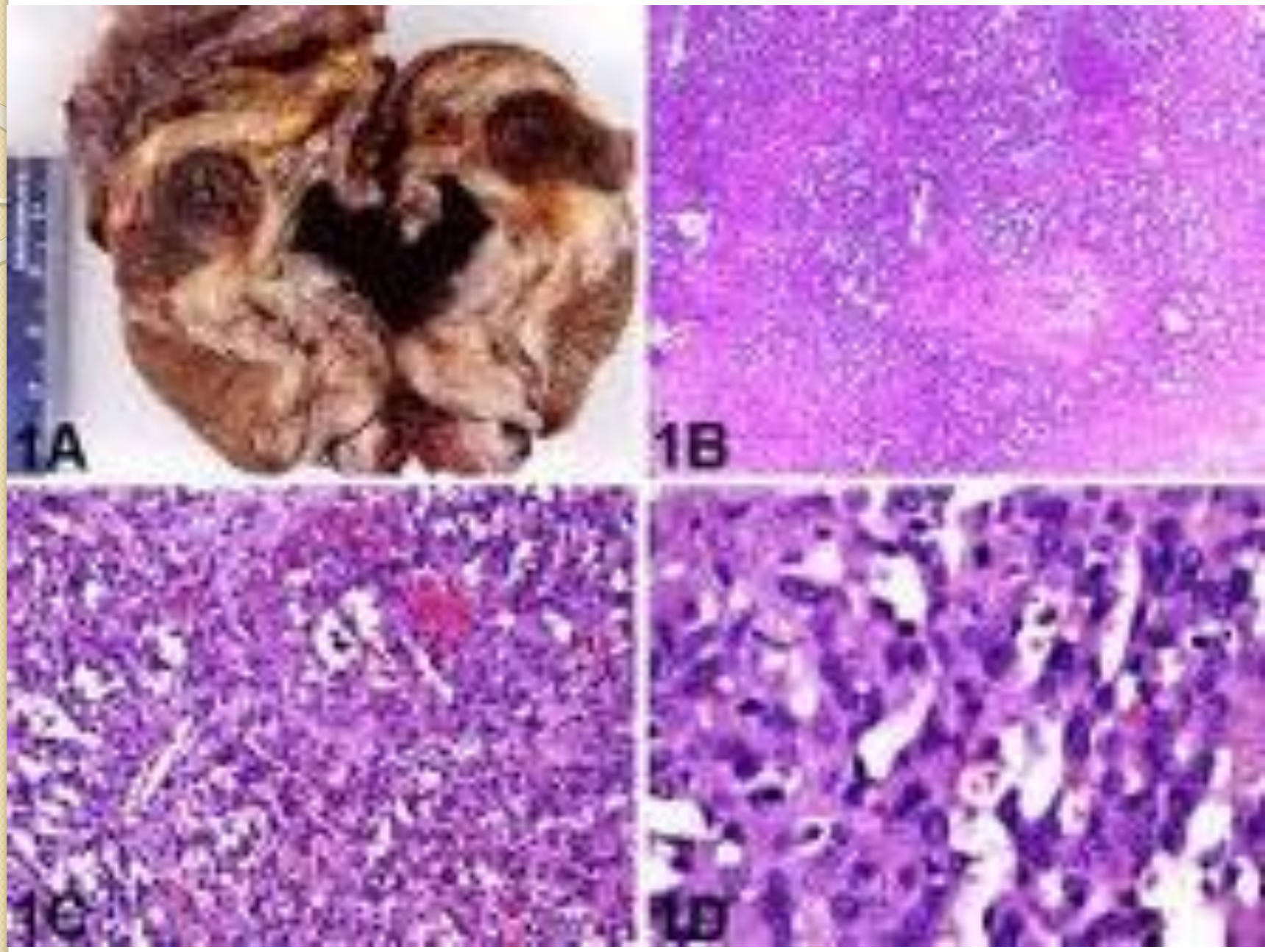
Angiosarcoma

- Malignant
- Highly differentiated tumors resembling capillary hemangioma to undifferentiated tumors resembling carcinomas or melanomas
- Old age
- More commonly in skin, soft tissue, breast and liver

- Hepatic angiosacomas are associated with distinct carcinogens, including arsenic, thorotrast & polyvinyl chloride
- In the setting of lymphedema after mastectomy
- Induced by radiation also

Morphology

- Grossly cutaneous angiosarcoma may begin as small asymptomatic lesion.
- Eventually most of them become large, fleshly masses of pale soft tissue
- All degrees of differentiation are identified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Thank you