



The Heart

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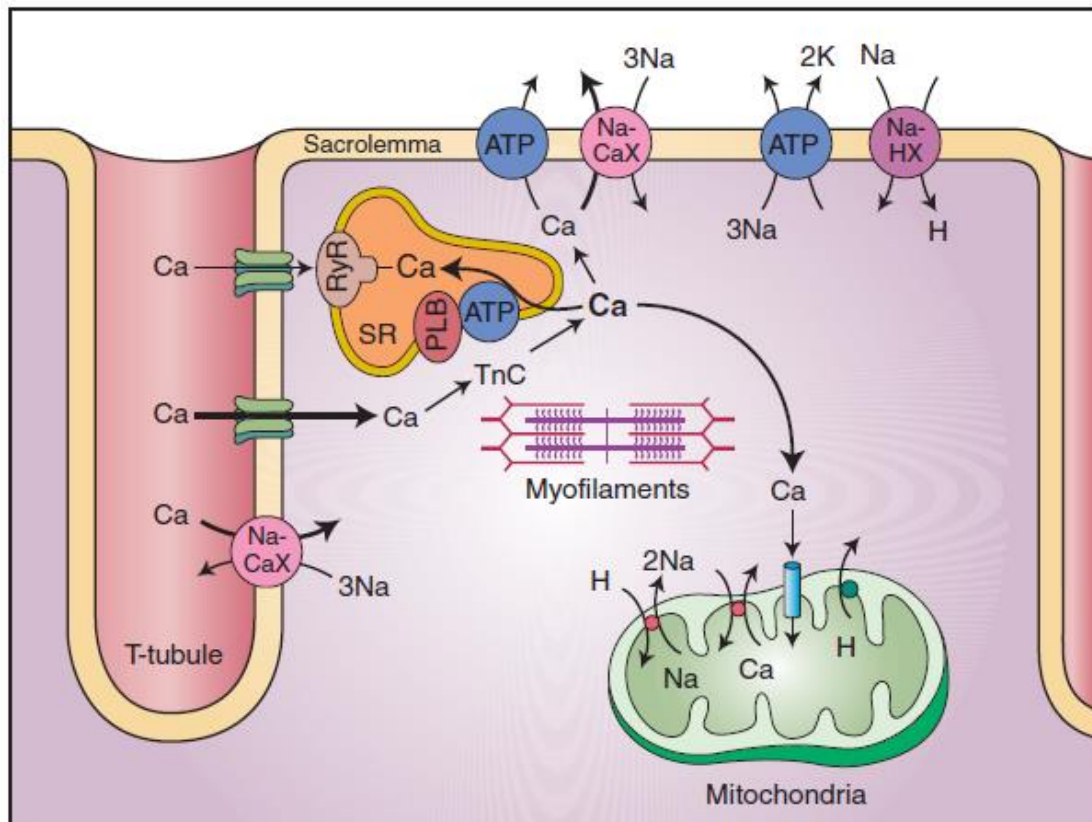
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Anatomy of the Heart

®The heart of an adult man weighs 280 to 340 g, and that of a woman, 230 to 280 g. Blood enters each side through a thin-walled atrium, from which it is propelled forward by thicker muscular ventricles. The right ventricle is considerably thinner (0.5 cm) than the left ventricle (1.3 to 1.5 cm) owing to the low venous pressure and relatively low afterload on the right side. The heart wall has three layers: outer epicardium, middle myocardium and inner endocardium. The heart is surrounded and enclosed by visceral and parietal pericardia, which are separated by the pericardial cavity.

Contraction of cardiac muscle is initiated by increases in cytosolic free calcium and the contractile force of the heart is a function of fiber length during diastole.

The cardiac action potential brings depolarizing current into T tubules where voltage-gated L-type Ca^{2+} channels reside in high concentrations (green channel structures). Influx of Ca^{2+} through these channels (I_{Ca}) stimulates release of Ca^{2+} from the sarcoplasmic reticulum (SR) (located in immediate proximity to the T tubule) via cardiac ryanodine receptor (RyR2). The transient increase in cytosolic Ca^{2+} promotes contraction through interactions with cardiac troponin T (TnC). Resting diastolic Ca^{2+} levels are restored by reuptake into the SR and extrusion via sodium–calcium exchange (Na-CaX) and an adenosine triphosphate (ATP) pump.

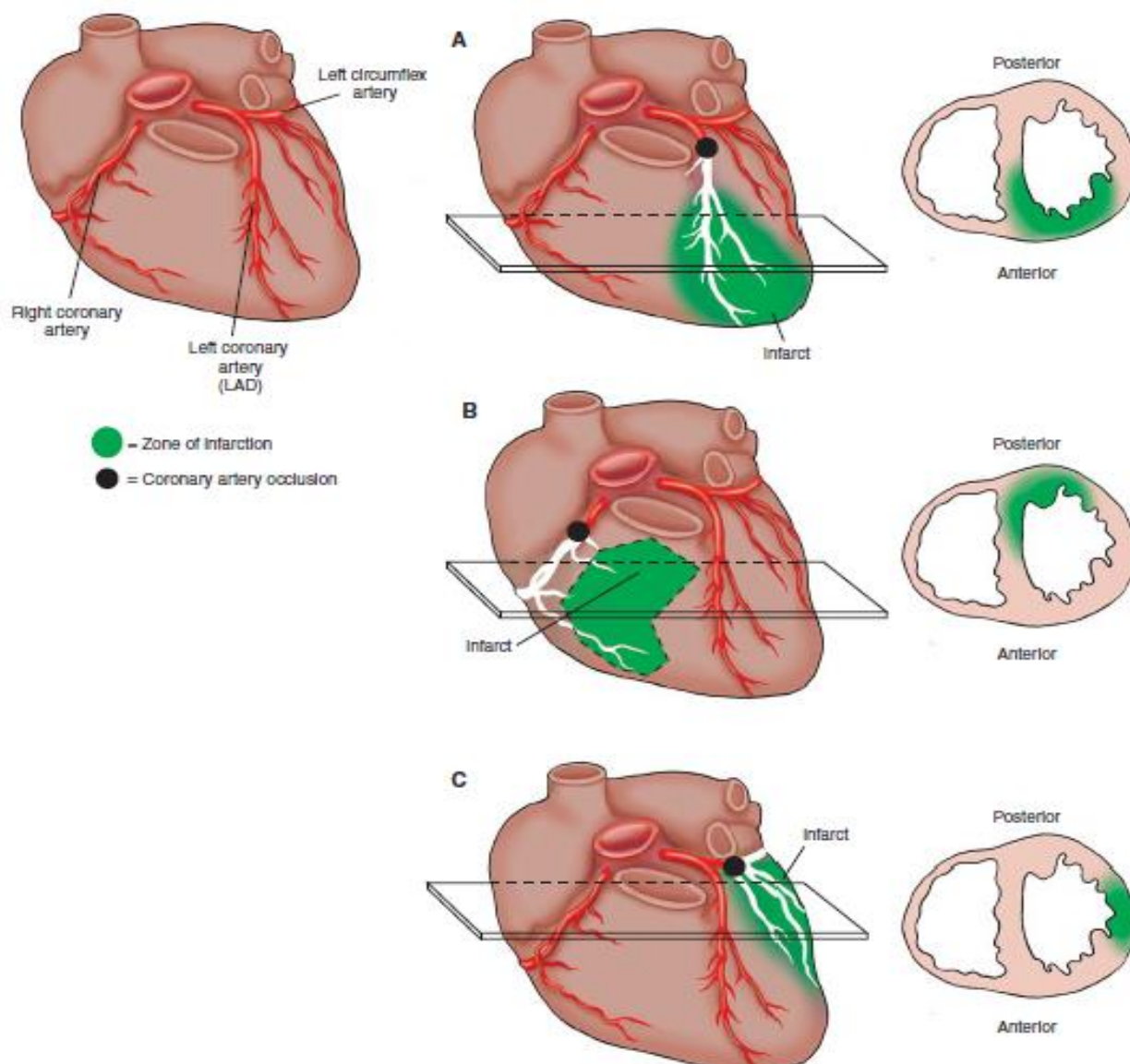


Coronary Arteries Supply Blood to the Heart

The right and left main coronary arteries originate in, or immediately above, the sinuses of Valsalva of the aortic valve.

The left main coronary artery bifurcates within 1 cm of its origin into the left anterior descending (LAD) and left circumflex coronary arteries. The left circumflex coronary artery rests in the left atrioventricular groove and supplies the lateral wall of the left ventricle. The LAD coronary artery lies in the anterior interventricular groove and provides blood to the (1) anterior left ventricle, (2) adjacent anterior right ventricle and (3) anterior half to two thirds of the interventricular septum. In the apical region, the LAD artery supplies the ventricles circumferentially.

The right coronary artery travels along the right atrioventricular groove and nourishes the bulk of the right ventricle and posteroseptal left ventricle, including the posterior third to half of the interventricular septum at the base of the heart (also referred to as the “inferior” or “diaphragmatic” wall). From these distributions, one can predict the location of infarcts that result from occlusion of any of the three major epicardial coronary arteries.



Myocardial Hypertrophy and Heart Failure

When a heart is injured, the clinical consequences are similar, regardless of the cause of cardiac dysfunction. If the initial impairment is severe, cardiac output is not maintained despite compensatory changes and the result is acute, lifethreatening, cardiogenic shock. When the functional impairment is less, compensatory mechanisms maintain cardiac output by increasing diastolic ventricular filling pressure and end-diastolic volume. This situation results in the characteristic signs and symptoms of congestive heart failure.

Hypertrophy initially reflects compensatory and potentially reversible mechanisms, but in the face of persistent stress, the myocardium becomes irreversibly enlarged and dilated. Ventricular hypertrophy is seen in virtually all conditions associated with chronic heart failure.

Left-sided heart failure is more common, because the most frequent causes of cardiac injury (e.g., ischemic heart disease and hypertension) primarily affect the left ventricle.

Right-sided heart failure commonly complicates leftsided failure, or it can develop independently secondary to intrinsic pulmonary disease or pulmonary hypertension, which creates resistance to blood flow through the lungs.

Diastolic heart failure is often seen in elderly patients. Ventricles become progressively stiffer with advancing age, and require greater filling (diastolic) pressures. These patients do not tolerate increases in blood volume well and are susceptible to developing pulmonary edema in response to a fluid challenge.

Congenital Heart Disease (CHD)

(Classifications of Congenital Heart Disease Reflect Cyanosis and Shunting)

Three groups based on the presence or absence of cyanosis

The acyanotic group does not have an abnormal communication between the systemic and pulmonary circuits. Examples of the acyanotic group include coarctation of the aorta, right-sided aortic arch and Ebstein malformation.

The cyanose tardive group is defined as an initial left-to-right shunt with late reversal of flow, including patent ductus arteriosus (PDA), patent foramen ovale and ventricular septal defect. In patients with these anomalies, cyanosis supervenes later (i.e., tardive). Although the shunt is initially left to right, it later becomes right to left (Eisenmenger complex) because progressive increases in pulmonary vascular resistance cause the right ventricular pressure to rise to the point where it exceeds that in the left ventricle

The cyanotic group describes a permanent right-to-left shunt. This category of CHD includes tetralogy of Fallot, truncus arteriosus, tricuspid atresia and complete transposition of the great vessels.

(Early Left-to-Right Shunt Reflects the Higher Pressure on the Left Side of the Heart)

Ventricular Septal Defect

VSDs occur as (1) a small hole in the membranous septum; (2) a large defect involving more than the membranous region (perimembranous defects); (3) defects in the muscular portion, which are more common anteriorly but can occur anywhere in the muscular septum and are often multiple; or (4) complete absence of the muscular septum (leaving a single ventricle).

Atrial Septal Defects

Patent foramen ovale: Tissue derived from the septum primum situated on the left side of the foramen ovale functions as a flap valve that normally fuses with the margins of the foramen ovale, thereby sealing the opening.

Atrial septal defect, ostium secundum type: defect occurs in the middle portion of the septum and varies from a trivial opening to a large defect of the entire fossa ovalis region.

Sinus venosus defect: This anomaly, accounting for 5% of ASDs, occurs in the upper portion of the atrial septum, above the fossa ovalis, near the entry of the superior venacava

Atrial septal defect, ostium primum type: This condition involves the region adjacent to the endocardial cushions. There are usually clefts in the anterior leaflet of the mitral valve and the septal leaflet of the tricuspid valve, which may be accompanied by a defect in the adjacent interventricular septum.

Atrioventricular canal:

Coronary sinus atrial septal defect: This is the rarest of the atrial septal defects. It is situated in the posteroinferior part of the interatrial septum at the site of the coronary sinus ostium, and is associated with a persistent left superior vena cava, which drains into the roof of the left atrium.

Patent Ductus Arteriosus

The left sixth aortic arch is partly preserved as the pulmonary arteries, and the arterial continuation on the left to the descending thoracic aorta becomes the ductus arteriosus. The ductus conveys most of the pulmonary outflow into the aorta, but constricts after birth in response to the increased arterial oxygen content and becomes occluded by fibrosis (ligamentum arteriosus)

Persistent Truncus Arteriosus

The truncus arteriosus is the embryonic arterial trunk that initially opens from both ventricles and is later separated into the aorta and the pulmonary trunk by the spiral septum. Persistent truncus arteriosus is a common trunk of origin for the aorta, pulmonary arteries and coronary

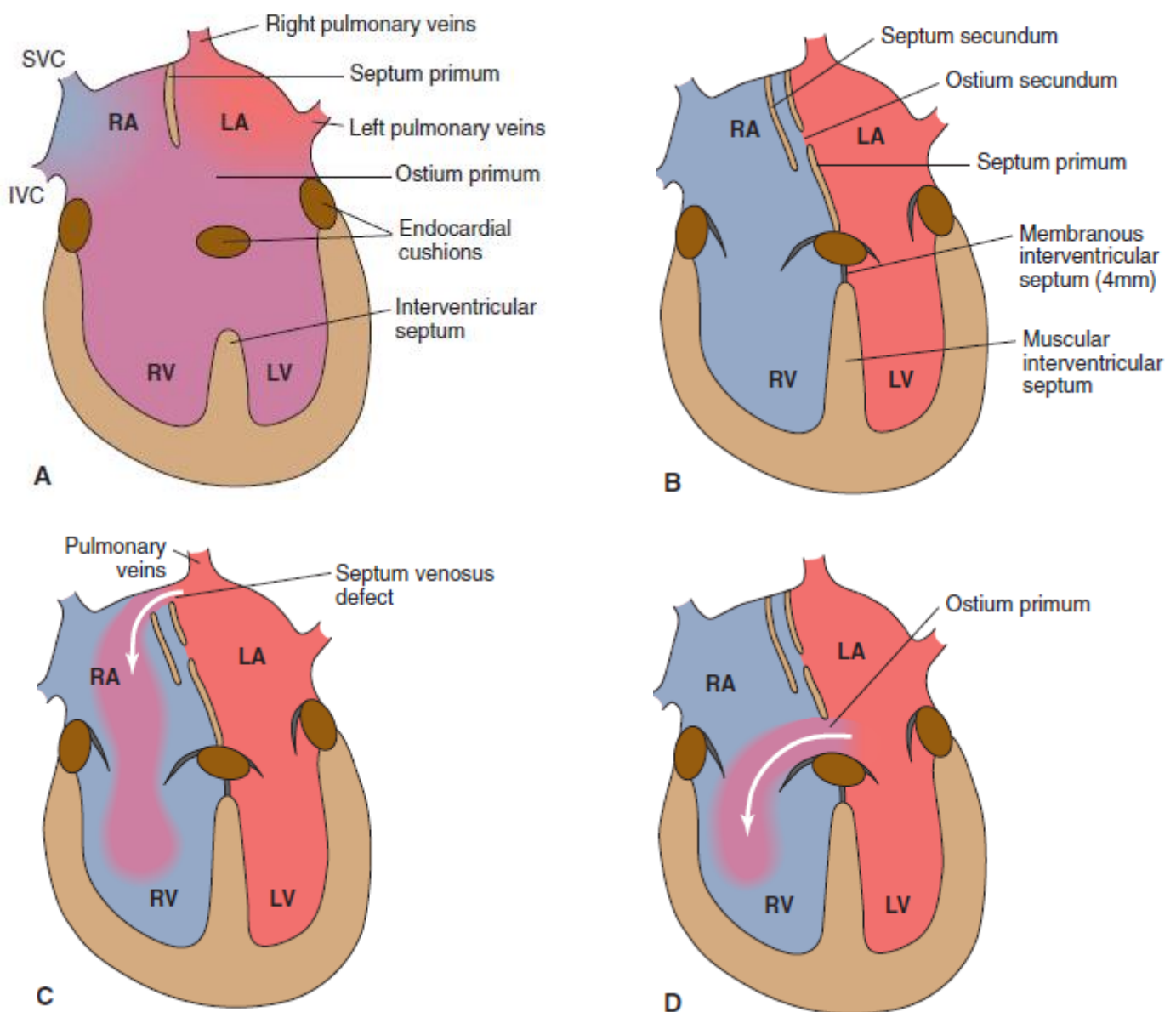
arteries, resulting from absent or incomplete partitioning of the truncus arteriosus by the spiral septum. Truncus arteriosus always overrides a VSD and receives blood from both ventricles.

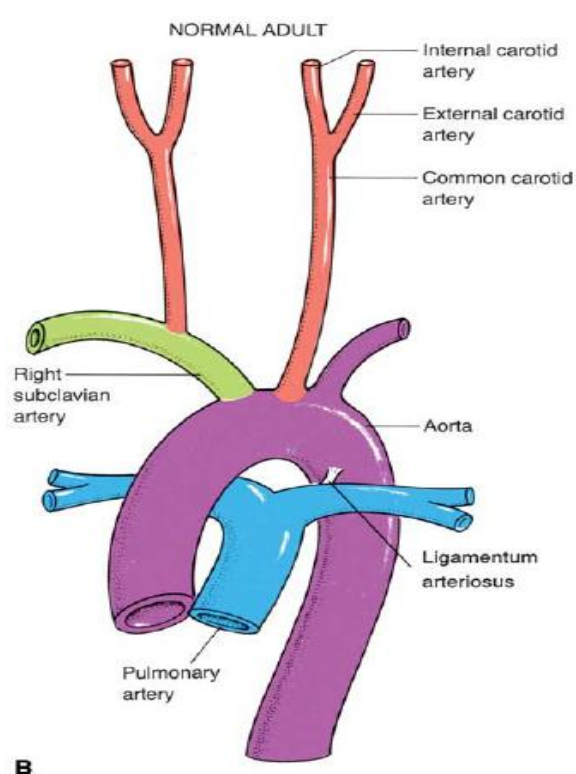
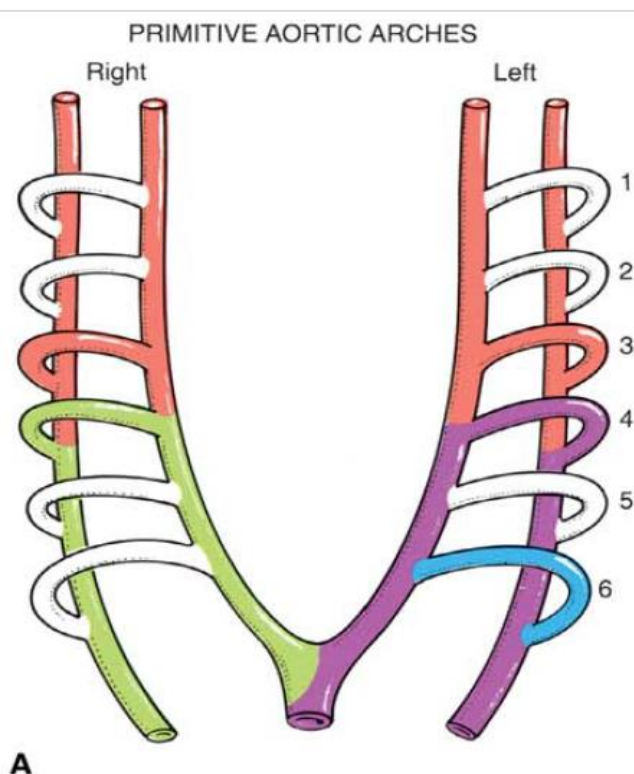
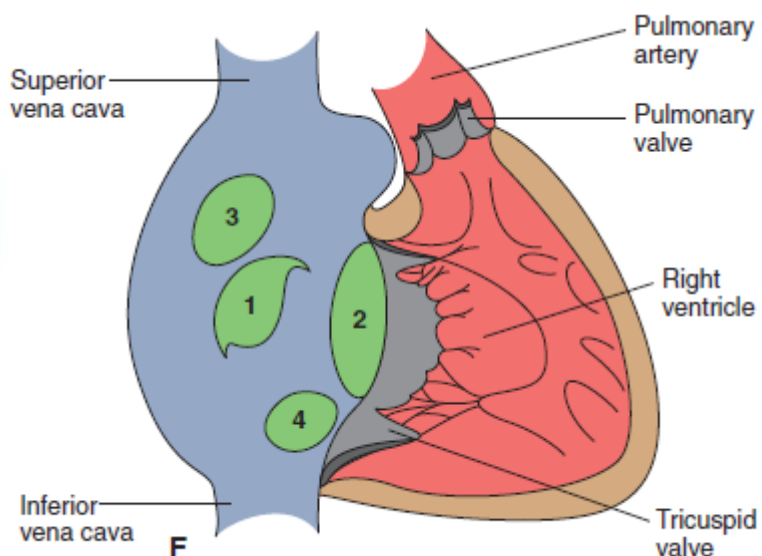
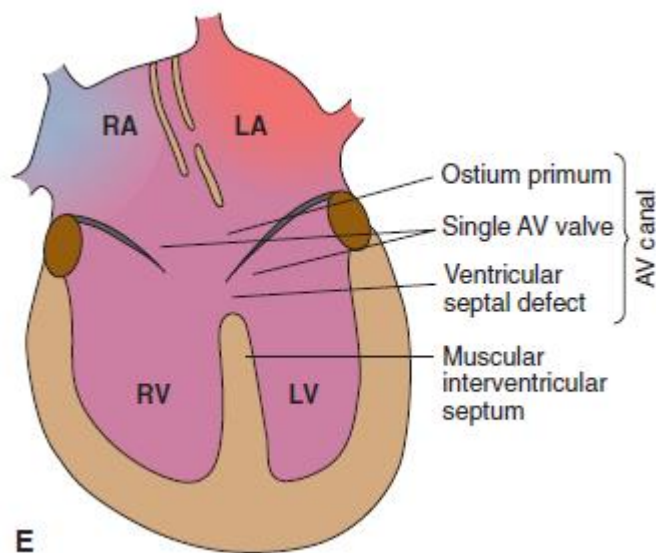
Hypoplastic Left Heart Syndrome

This usually profound malformation is characterized by hypoplasia of the left ventricle and ascending aorta and hypoplasia or atresia of the left-sided valves.

Anomalous Pulmonary Vein Drainage

The pulmonary veins form a network in the dorsal mesoderm. A bud from the region of the atrium joins the pulmonary venous confluence, and eventually all four pulmonary veins drain into the left atrium. Failure of these tissues to join correctly results in various venous anomalies.





(Right-to-Left Shunt Is the Most Common Cyanotic Congenital Heart Disease)

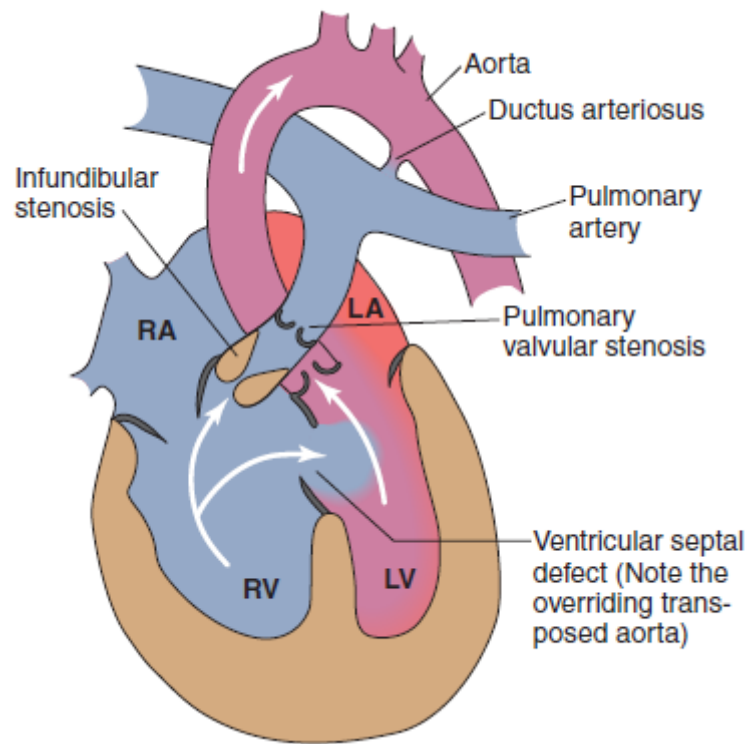
Tetralogy of Fallot

The four anatomic changes that define the tetralogy of Fallot are

- Pulmonary stenosis
- Ventricular septal defect
- Dextroposition of the aorta so that it overrides the ventricular septal defect
- Right ventricular hypertrophy

Tricuspid Atresia

Tricuspid atresia, a congenital absence of the tricuspid valve, results in an obligate right-to-left shunt through the patent foramen ovale.

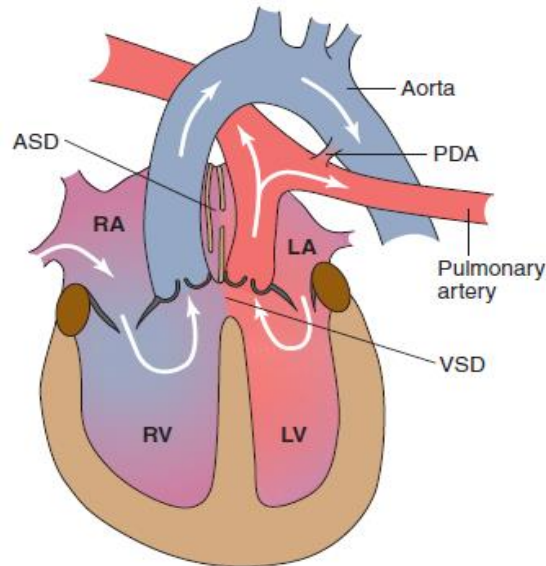


Tetralogy of Fallot. Note the pulmonary stenosis, which is due to infundibular hypertrophy as well as pulmonary valvular stenosis. The ventricular septal defect involves the membranous septum region. Dextroposition of the aorta and right ventricular hypertrophy are shown. Because of the pulmonary obstruction, the shunt is from right to left, and the patient is cyanotic. LA _ left atrium; LV _ left ventricle; RA _ right atrium; RV _ right ventricle.

(Congenital Heart Diseases Without Shunts Involve Various Cardiovascular Sites)

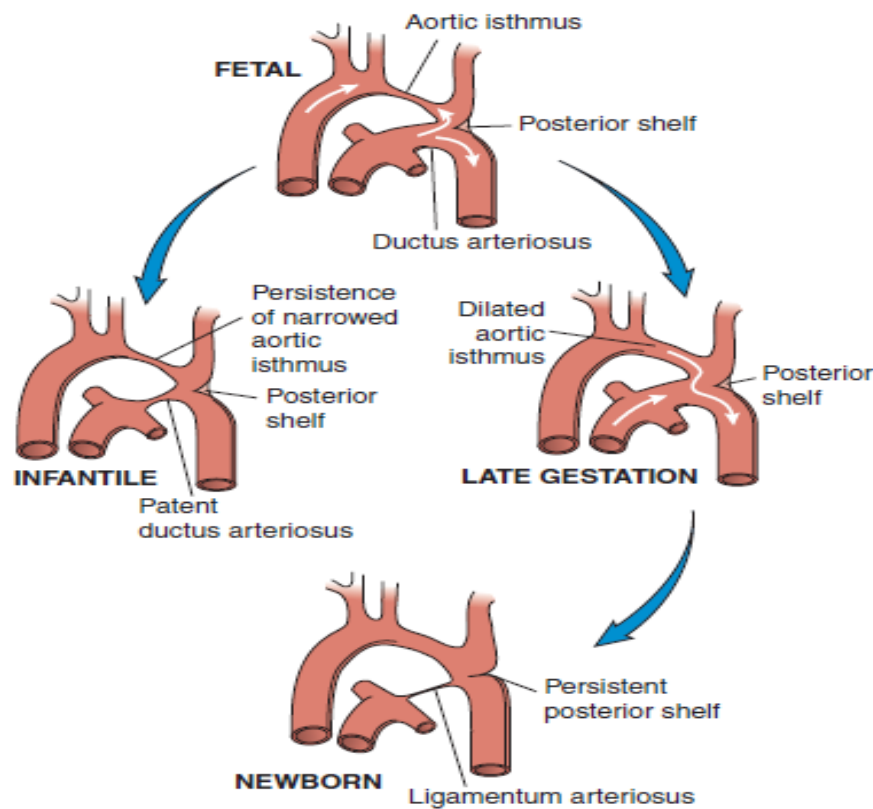
Transposition of the Great Arteries

In transposition of the great arteries (TGA), the aorta arises from the right ventricle and the pulmonary artery from the left ventricle.



Coarctation of the Aorta

Coarctation of the aorta is a local constriction that almost always occurs immediately below the origin of the left subclavian artery at the site of the ductus arteriosus.



Pathogenesis of coarctation of the aorta. In the fetus, ductal blood is diverted into cephalad and descending streams by the posterior aortic shelf. In late fetal life, the isthmus dilates and the increased descending blood flow is accommodated by the ductal orifice. After birth, if the shelf does not undergo the normal involution, obliteration of the ductal orifice does not permit free flow around the persistent posterior shelf, thereby creating a juxtaductal obstruction of blood flow to the distal aorta. If the aortic isthmus does not dilate during late fetal life, it remains narrow, resulting in an infantile or preductal coarctation. In this circumstance, the ductus arteriosus usually remains patent.

Pulmonary Stenosis

Pulmonary stenosis results from (1) developmental deformities arising from the endocardial cushion region of the heart (with involvement of the pulmonary valves), (2) an abnormality of the right ventricular infundibular muscle (subvalvular or infundibular stenosis, especially as part of tetralogy of Fallot) or (3) abnormal development of the more distal parts of the pulmonary artery tree (peripheral pulmonary stenosis).

Congenital Aortic Stenosis

Valvular Aortic Stenosis: The most common congenital aortic stenosis, bicuspid valve, arises through abnormal development of the endocardial cushions.

-Subvalvular Aortic Stenosis

-Supravalvular Aortic Stenosis

-Origin of a Coronary Artery From the Pulmonary Artery

-Ebstein Malformation

-Congenital Heart Block

-Endocardial Fibroelastosis

-Dextrocardia

Ischemic Heart Disease

Ischemic heart disease is, in most cases, a consequence of coronary artery atherosclerosis. It develops when blood flow is inadequate to meet the oxygen demands of the heart.

ANGINA PECTORIS: This term refers to the pain of myocardial ischemia. It typically produces a severe crushing or burning sensation in the substernal portion of the chest and may radiate to the left arm, jaw or epigastrium. It is the most common symptom of ischemic heart disease.

Prinzmetal angina (variant angina) is an atypical form of angina that occurs at rest and is caused by coronary artery spasm.

Unstable angina, a variety of chest pain that has a less predictable relationship to exercise than does stable angina and may occur during rest or sleep, is associated with development of nonocclusive thrombi over atherosclerotic plaques.

MYOCARDIAL INFARCT: A myocardial infarct is a discrete focus of ischemic muscle necrosis in the heart.

CHRONIC CONGESTIVE HEART FAILURE: Because early mortality associated with acute myocardial infarction is now less than 5%, many patients with ischemic heart disease survive longer and eventually develop chronic congestive heart failure. Coronary artery disease is responsible for heart failure in more than 75% of all patients with heart failure.

SUDDEN DEATH: In some patients, the first and only clinical manifestation of ischemic heart disease is sudden death due to spontaneous ventricular tachycardia that degenerates into ventricular fibrillation. In any event, coronary atherosclerosis underlies most cases of cardiac death occurring during the first hour after the onset of symptoms.

Risk factors for ischemic heart disease include

Obesity, Age, Sex (Sixty percent of coronary events occur in men), **Family history, Use of oral contraceptives, Sedentary life habits, Personality features.**

Conditions Limit the Supply of Blood to the Heart

1-Atherosclerosis and Thrombosis

2-Thromboemboli

3-Coronary Collateral Circulation (most normal hearts have anastomoses 20 to 200 μ m in diameter between coronary vessels, these collateral vessels do not function under normal circumstances because there is no pressure gradient between the arteries that they connect.)

4-Other Conditions (Coronary arteritis, Dissecting aneurysm of the aorta, Syphilitic aortitis, Congenital anomalous origin of a coronary artery, An intramural course of the LAD coronary artery)

Blood to Deliver Oxygen Is Limitation lead to Myocardium Risk for Ischemia

Anemia is a common cause of decreased oxygen delivery to the myocardium.

Carbon monoxide (CO) poisoning decreases oxygen delivery to the tissues.

Increased Oxygen Demand May Cause Cardiac Ischemia

Any increase in cardiac workload increases the heart's need for oxygen. Conditions that raise blood pressure or cardiac output, such as exercise or pregnancy, augment oxygen demand by the myocardium

Types Myocardial Infarction (Subendocardial or Transmural)

-A subendocardial infarct affects the inner one third to one half of the left ventricle, generally results from hypoperfusion of the heart. It may arise within the territory of one of the major **epicardial coronary arteries**

-A transmural infarct involves the full left ventricular wall thickness, usually after occlusion of a coronary artery. As a result, transmural infarcts typically conform to the distribution of **one of the three major coronary arteries**.

Complications of Myocardial Infarction:

- 1-Early mortality in acute myocardial infarction
- 2- Arrhythmias
- 3- Left ventricular failure and cardiogenic shock
- 4- Extension of the infarct
- 5- Rupture of the free wall of the myocardium
- 6- Aneurysms
- 7- Mural thrombosis and embolism
- 8- Pericarditis

Chronic Ischemic Heart Disease CanLead to Cardiomyopathy

In a minority of patients with severe coronary atherosclerosis, myocardial contractility is impaired globally without discrete infarcts, as in dilated cardiomyopathy.

Hypertensive Heart Disease

Hypertension has been defined as a persistent increase of systemic blood pressure above 140 mm Hg systolic or 90 mm Hg diastolic, or both, Chronic hypertension leads to pressure overload resulting first in compensatory left ventricular hypertrophy and, eventually, cardiac failure. The term hypertensive heart disease is used when the heart is enlarged in the absence of a cause other than hypertension. Diastolic dysfunction is the most common functional abnormality caused by hypertension and by itself can lead to congestive heart failure.

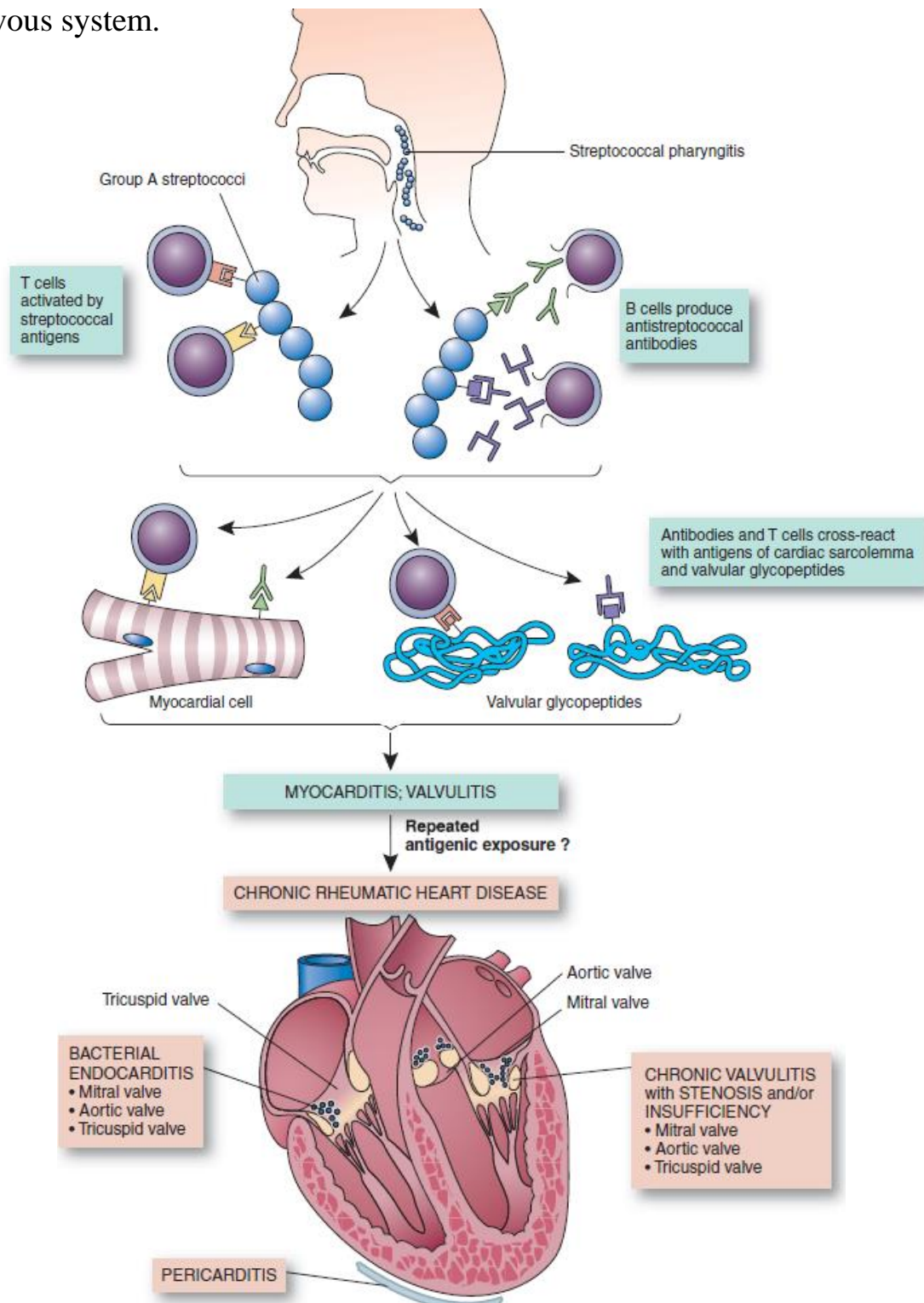
Cor Pulmonale

Cor pulmonale is right ventricular hypertrophy and dilation due to pulmonary hypertension.

Acquired Valvular and Endocardial Diseases

A variety of inflammatory, infectious and degenerative diseases damage cardiac valves and impair their function. When valves become damaged, leaflets or cusps may be thickened and fused enough to narrow the aperture and obstruct blood flow, a condition labeled **valvular stenosis**, and diseases that destroy valve tissue may also allow retrograde blood flow, termed **valvular regurgitation or insufficiency**.

Rheumatic fever (RF) is a multisystem childhood disease that follows a streptococcal infection and is characterized by an inflammatory reaction involving the heart, joints and central nervous system.



Collagen Vascular Diseases Affect Both Cardiac Valves and Myocardium

-Systemic Lupus Erythematosus

The most common cardiac lesion is fibrinous pericarditis, usually with an effusion. Myocarditis, at least in the form of subclinical left ventricular dysfunction.

-Rheumatoid Arthritis

Pericardium, myocardium or valves involvement of the heart in rheumatoid arthritis does not compromise function.

- Ankylosing Spondylitis

The aortic valve ring is dilated and its cusps are scarred and shortened. Focal inflammatory lesions occur in all layers of the aortic wall, particularly near the valve ring.

- Scleroderma (Progressive Systemic Sclerosis)

Cardiac involvement is second only to renal disease as a cause of death in scleroderma. The myocardium exhibits intimal sclerosis of small arteries, which leads to small infarcts and patchy fibrosis.

-Polyarteritis Nodosa

Necrotizing lesions in branches of the coronary arteries result in myocardial infarction, arrhythmias or heart block.

Nonbacterial Thrombotic Endocarditis (NBTE) Is a Complication of Wasting Diseases

Also known as marantic endocarditis, refers to sterile vegetations on apparently normal cardiac valves, almost always in association with cancer or some other wasting disease.

Calcific Aortic Stenosis Reflects Chronic Damage to the Valve

Calcific aortic stenosis refers to narrowing of the aortic valve orifice due to calcium deposition in the valve cusps and ring.

Mitral Valve Prolapse (MVP) Is the Most Common Indication for Valve Repair or Replacement

MVP is a condition in which mitral valve leaflets become enlarged and redundant. Chordae tendineae become thinned and elongated, such that the billowed leaflets prolapse into the left atrium during systole

Papillary Muscle Dysfunction May Produce Mitral Regurgitation

The papillary muscles are especially vulnerable to ischemic injury because they are supplied by the terminal branches of the intramyocardial coronary arteries. Thus, any reduction in coronary blood flow may preferentially interfere with papillary muscle function.

Carcinoid Heart Disease Affects Right-Sided Valves

Carcinoid heart disease is an unusual condition that uniquely affects the right side of the heart, leading to tricuspid regurgitation and pulmonary stenosis.

Myocarditis:

Myocarditis is inflammation of the myocardium associated with myocyte necrosis and degeneration.

-**Viral** Myocarditis are without an easily demonstrable cause

-**Granulomatous** Myocarditis may be caused by microorganisms or immunologically mediated injury

- **Hypersensitivity** Myocarditis is a reaction to drugs

-**Giant Cell** Myocarditis is usually fatal (The cause is unknown, but it sometimes occurs in patients with SLE, hyperthyroidism or thymoma. An autoimmune etiology has been suggested, but there is no persuasive evidence for this theory.

Metabolic Diseases of the Heart

Hyperthyroidism Causes High-Output Failure

Thyroid hormone has direct inotropic and chronotropic effects on the heart **(1)** it increases the activity of the sarcolemmal sodium pump; **(2)** it enhances the synthesis of a myosin isoform with rapid ATPase activity and reduces production of a slower isoform; and **(3)** it upregulates expression of slow calcium channels in the sarcolemma, thereby facilitating contractility. **Hyperthyroidism thus causes** conspicuous tachycardia and an increased cardiac workload, owing to decreased peripheral resistance and increased cardiac output. It may eventually lead to angina pectoris and high-output failure.

Hypothyroid Heart Disease Diminishes Cardiac Output

There may be a pericardial effusion created by increased capillary permeability and leakage of fluid and protein into the pericardial cavity. Pulse pressure is decreased because of higher peripheral resistance and lower blood volume.

Thiamine Deficiency (Beriberi) Heart Disease Is Similar to Hyperthyroidism

Appear at diet consists largely of shelled rice or/and alcoholics or neglected persons.

Cardiomyopathy

Cardiomyopathy refers to a primary disease of the myocardium. strictly defined, it excludes damage caused by extrinsic factors. the primary cardiomyopathies are divided into the major clinicopathologic groups

Dilated cardiomyopathy (DCM) : characterized by impaired contractility, At autopsy, the heart is invariably enlarged, with conspicuous left and right ventricular hypertrophy.

Hypertrophic cardiomyopathy (HCM): Is out of Proportion to the Hemodynamic Load

Arrhythmogenic right ventricular cardiomyopathy (ARVC): Is a disease of the Desmosome with a high risk of sudden death

Restrictive cardiomyopathy (RCM): Impairs diastolic function, describes a group of diseases in which myocardial or endocardial abnormalities limit diastolic filling while contractile function remains normal.

Sudden Cardiac Death

Most of these deaths are caused by spontaneous lethal ventricular tachyarrhythmias ventricular tachycardia and ventricular fibrillation—in patients with some type of heart disease.

In economically developed nations, coronary artery disease is responsible for most sudden deaths in middle-aged and older adults, lethal arrhythmias usually arise from pathologic changes affecting conduction properties of the working ventricular myocardium, sudden cardiac death occurs in patients with structurally normal hearts, but this is rare

Cardiac Tumors

-**Cardiac Myxomas** are the most common primary tumors of the heart

-**Rhabdomyoma** Is the most common primary childhood cardiac tumor It may actually be a hamartoma rather than a true neoplasm

-**Papillary Fibroelastoma:** involves the valves

Diseases of the Pericardium

1-Pericardial Effusion: is accumulation of excess fluid within the pericardial cavity, as either a transudate or an exudate, it can cause cardiac Tamponade

Types of Pericardial Effusion:

*Serous pericardial effusion: The fluid has a low protein content and few cellular elements.

*Chylous effusion (fluid containing chylomicrons) results from a communication of the thoracic duct with the pericardial space due to lymphatic obstruction by tumor or infection.

* Serosanguineous pericardial effusion may develop after chest trauma, either accidentally or caused by cardiopulmonary resuscitation.

*Hemopericardium is bleeding directly into the pericardial cavity

2-Acute Pericarditis: Pericarditis refers to inflammation of the visceral or parietal pericardium may follow viral infections.

3-Constrictive Pericarditis: Constrictive pericarditis is a chronic fibrosing disease of the pericardium that compresses the heart and restricts inflow. may mimic right heart failure.

Pathology of Interventional Therapies

1-**Percutaneous Coronary Interventions:** are used to treat Atherosclerotic coronary disease

2-**Coronary Bypass Grafts:** circumvent obstructed segments

3- **Valve Replacements**

4- **Heart Transplantation:** may cure many end-stage heart diseases but Is subject to host rejection processes