allows the SA node to take control again, returning the heart to sinus rhythm. These devices may be external or implanted internally. Newer devices have electronic memory, which can be downloaded to assess cardiac function and efficiency of the device.

THINK ABOUT 12-8

a. Compare PVCs, atrial flutter, atrial fibrillation, and total heart block.
b. Using one type of dysrhythmia as an example, explain how cardiac output may be reduced.
c. Explain the absence of peripheral pulses in ventricular fibrillation.

Cardiac Arrest or Standstill (Asystole)

Cardiac arrest is the cessation of all activity in the heart. There is no conduction of impulses, and the ECG shows a flat line. Lack of contractions means that no cardiac output occurs, thus depriving the brain and heart itself of oxygen. Loss of consciousness takes place immediately, and respiration ceases. There is no pulse at any site, including the apical and carotid sites (see Fig. 12-17).

Arrest may occur for many reasons; for example, excessive vagal nerve stimulation may slow the heart, drug toxicity may occur, or there may be insufficient oxygen to maintain the heart tissue due to severe shock or ventricular fibrillation. In order to resuscitate a person, blood flow to the heart and brain must be maintained.

EMERGENCY TREATMENT FOR CARDIAC ARREST

1. Call for emergency medical help and begin CPR.
2. Commence use of an automatic electrical defibrillator if one is available. (These are located in public buildings and marked with a red symbol showing an electrical flash through a heart. The letters AED appear on the cover [Fig. 12-19].)
3. Continue CPR if no AED device is available or if instructed to do so by the device.

FIGURE 12-19 The universal AED symbol indicates presence and location of automatic electrical defibrillator. Symbol may be red or green in color.

Congestive Heart Failure

Pathophysiology

Congestive heart failure occurs when the heart is unable to pump sufficient blood to meet the metabolic needs of the body. Usually CHF occurs as a complication of another condition. It may present as an acute episode but usually is a chronic condition. It may result from a problem in the heart itself, such as infarction or a valve defect; it may arise from increased demands on the heart, such as those imposed by hypertension or lung disease; or it may involve a combination of factors. Depending on the cause, one side of the heart usually fails first, followed by the other side. For example, an infarction in the left ventricle or essential hypertension (high blood pressure) affects the left ventricle first, whereas pulmonary valve stenosis or pulmonary disease affects the right ventricle first. It is helpful in the early stages to refer to this problem as left-sided CHF or right-sided CHF.

Initially various compensation mechanisms maintain cardiac output (Fig. 12-20, top part). Unfortunately, these mechanisms often aggravate the condition instead of providing assistance:

- The reduced blood flow into the systemic circulation and thus the kidneys leads to increased renin and aldosterone secretion. The resulting vasoconstriction (increased afterload) and increased blood volume (increased preload) add to the heart's workload.
- The SNS response also increases heart rate and peripheral resistance. Increased heart rate may decrease the efficiency of the heart and impede filling, as well as increasing work for the heart.
- The chambers of the heart tend to dilate (enlarge), and the cardiac muscle becomes hypertrophied (**cardiomegaly**), with the wall of the ventricle becoming thicker. This process demands increased blood supply to the myocardium itself, and eventually some myocardial cells die, to be replaced with fibrous tissue.

There are two basic effects when the heart cannot maintain its pumping capability:

1. *Cardiac output or stroke volume decreases,* resulting in less blood reaching the various organs and tissues, a "forward" effect. This leads to decreased cell function, creating fatigue and lethargy. Mild acidosis develops, which is compensated for by increased respirations (see Chapter 2). Because the affected ventricle cannot pump its load adequately, the return of blood to that side of the heart is also impaired.
2. *"Backup" congestion develops* in the circulation behind the affected ventricle (Fig. 12-21). The output from the ventricle is *less* than the inflow of blood.

For example, if the left ventricle cannot pump all of its blood into the systemic circulation, the normal volume of blood returning from the lungs cannot enter the left side of the heart. This eventually causes congestion in the pulmonary circulation, increased capillary

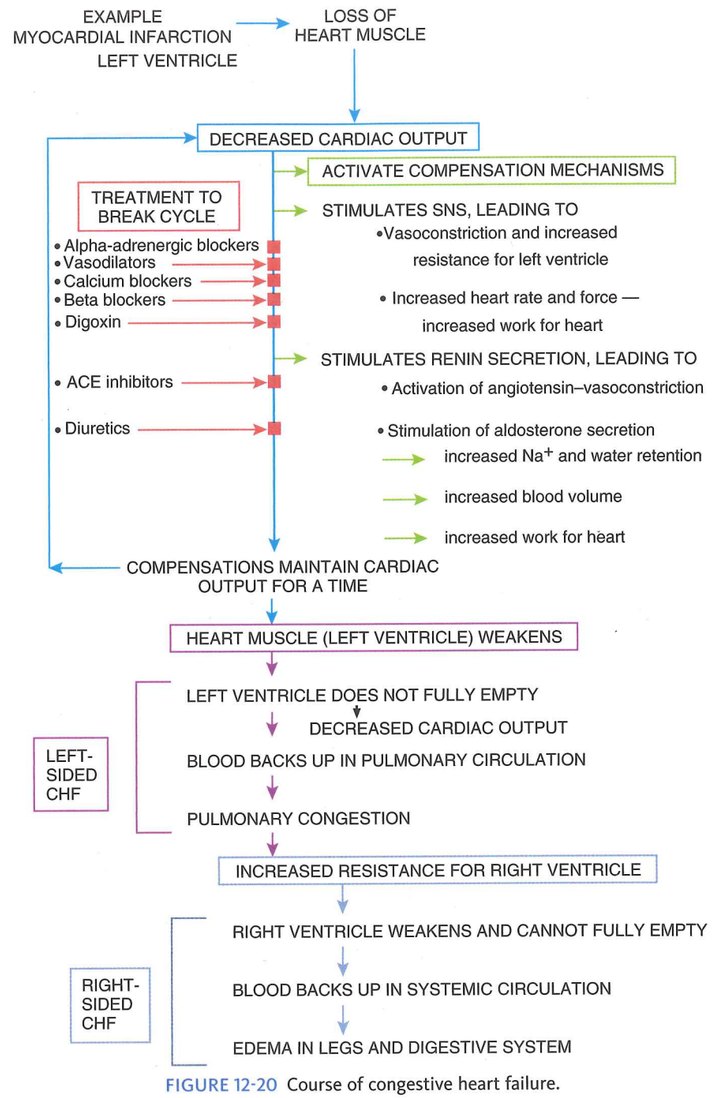

FIGURE 12-20 Course of congestive heart failure.

pressure, and possible pulmonary edema, in which fluid is forced into the alveoli. This situation is termed left-sided CHF.

In right-sided CHF, the right ventricle cannot maintain its output, so less blood proceeds to the left side of the heart and the systemic circulation (forward effect). The backup effect, or congestion, is apparent in the systemic circulation, as shown by increased blood volume and congestion in the legs and feet and eventually also in the portal circulation (liver and digestive tract) and neck veins. Right- and left-sided cardiac failures are compared in Table 12-3.

Etiology

Infarction that impairs the pumping ability or efficiency of the conducting system, valvular changes, or congenital heart defects may cause failure of the affected side. Presently coronary artery disease is the leading cause of CHF. Increased demands on the heart cause heart failure that may take various forms, depending on the ventricle most adversely affected. For example, essential hypertension increases diastolic blood pressure, requiring the left ventricle to contract with more force to open the aortic valve and eject blood into the aorta. The left ventricle hypertrophies and eventually

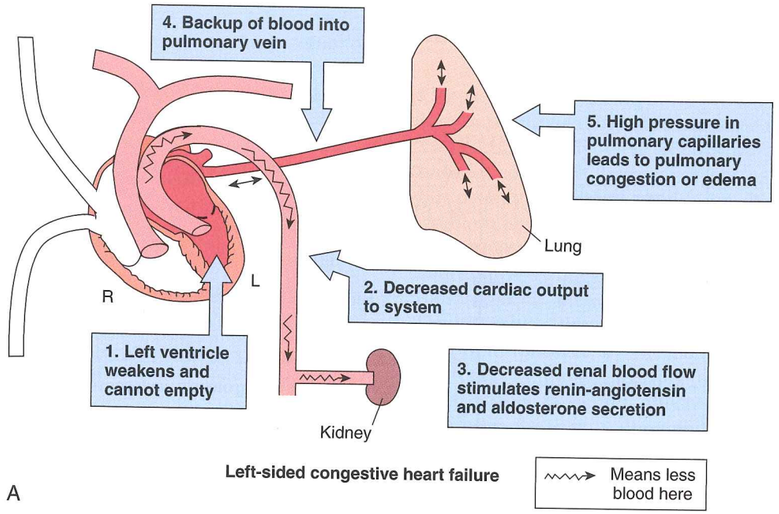

FIGURE 12-21 Effects of congestive heart failure.

fails (Fig. 12-22*A*). Pulmonary disease, which damages the lung capillaries and increases pulmonary resistance, increases the workload for the right ventricle; the muscle hypertrophies and eventually fails. Right-sided CHF due to pulmonary disease is often referred to as *cor pulmonale* (see Fig. 12-22*B* and further discussion in Chapter 13).

Signs and symptoms

The signs and symptoms become more marked as the condition progresses. Drugs may be controlling the severity of the manifestations, but there is an increased risk of sudden death from CHF.

1. With failure of either side, the *forward* effects are similar: decreased blood supply to the tissues and general hypoxia. Fatigue and weakness, dyspnea (breathlessness), and shortness of breath, especially with exertion, exercise intolerance, cold intolerance, and dizziness occur.
2. *Compensation* mechanisms are indicated by tachycardia, pallor, and daytime oliguria.
3. The *backup* effects of *left-sided* failure are related to pulmonary congestion and include:
   - Dyspnea and **orthopnea**, or difficulty in breathing when lying down, develop as increased fluid accumulates in the lungs in the recumbent position.
   - Cough is commonly associated with the fluid irritating the respiratory passages. The lungs become a dependent area when the body is recumbent. In this position as well, excess interstitial fluid returns to the blood, reducing edema but increasing blood volume and pooled fluid in the lungs.
   - *Paroxysmal nocturnal dyspnea* indicates the presence of *acute pulmonary edema.* This usually develops during sleep, when the increased blood volume in the lungs leads to increased fluid in the alveoli and interferes with oxygen diffusion and lung expansion. The individual awakes in a panic, struggling for air and coughing, sometimes producing a frothy, blood-stained sputum (**hemoptysis**) if capillaries have ruptured with the pressure.

TABLE 12-3 Congestive Heart Failure

	Left-Sided CHF	Right-Sided CHF
Causes		
	Infarction of left ventricle, aortic valve stenosis, hypertension, hyperthyroidism	Infarction of right ventricle, pulmonary valve stenosis, pulmonary disease (cor pulmonale)
Basic Effects		
	Decreased cardiac output, pulmonary congestion	Decreased cardiac output, systemic congestion, and edema of legs and abdomen
Signs and Symptoms		
Forward effects (decreased output)	Fatigue, weakness, dyspnea, exercise intolerance, cold intolerance	Fatigue, weakness, dyspnea, exercise intolerance, cold intolerance
Compensations	Tachycardia and pallor, secondary polycythemia, daytime oliguria	Tachycardia and pallor, secondary polycythemia, daytime oliguria
Backup effects	Orthopnea, cough producing white or pink-tinged phlegm, shortness of breath, paroxysmal nocturnal dyspnea, hemoptysis, rales	Dependent edema in feet, hepatomegaly and splenomegaly, ascites, distended neck veins, headache, flushed face

CHF, Congestive Heart Failure.

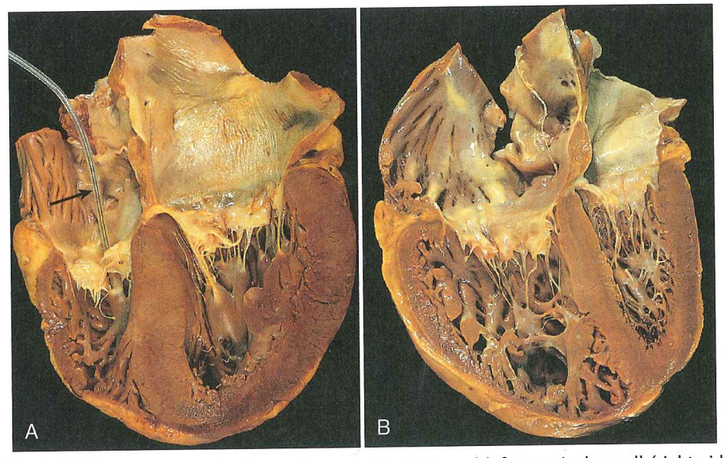

FIGURE 12-22 **A,** Hypertensive heart disease with thickened left ventricular wall (right side). Arrow indicates incidental pacemaker in the right ventricle. **B,** Chronic cor pulmonale showing dilated and enlarged right ventricle with thickened wall (left side). *(From Kumar V, Abbas AK, Fausto M:* Robbins and Cotran Pathologic Basis of Disease, *ed 7, Philadelphia, 2005, Saunders.)*

Rusty-colored sputum may be present with recurrent pulmonary edema, indicating the presence of hemosiderin-containing macrophages in the lungs. Rales (bubbly sounds of fluid in the lungs) and a rapid, weak pulse, together with cool, moist skin, are usually present. Sleeping with the upper body elevated may prevent this complication. Excess fluid in the lungs frequently leads to infections such as pneumonia.

4. Signs of *right-sided* failure and *systemic backup* include:
   - Dependent edema in the feet or legs or areas such as buttocks
   - Hepatomegaly and splenomegaly, and eventually digestive disturbances as the wall of the digestive tract becomes edematous
   - *Ascites*, a complication that occurs when fluid accumulates in the peritoneal cavity, leading to marked abdominal distention; hepatomegaly and ascites may impair respiration if upward pressure on the diaphragm impairs lung expansion
   - Acute right-sided failure, indicated by increased pressure in the superior vena cava, resulting in flushed face, distended neck veins, headache, and visual disturbances; this condition requires prompt treatment to prevent brain damage due to reduced perfusion of brain tissue.

Young Children with Congestive Heart Failure

Infants and young children manifest heart failure somewhat differently than adults. Heart failure is often secondary to congenital heart disease (see next section, Congenital Heart Defects). Feeding difficulties are often the first sign, with failure of the child to gain weight or meet developmental guidelines. Sleep periods are short because the baby falls asleep while feeding and is irritable when awake. There may be a cough, rapid grunting respirations, flared nostrils, and wheezing. With right-sided failure, hepatomegaly and ascites are common. Often a third heart sound is present (gallop rhythm).

Diagnostic Tests

Radiographs show cardiomegaly and the presence or absence of fluid in the lungs. Cardiac catheterization can be used to monitor the hemodynamics or pressures in the circulation. Arterial blood gases are used to measure hypoxia.

Treatment

The underlying problem should be treated if possible. Reducing the workload on the heart by avoiding excessive fatigue, stress, and sudden exertion is important in preventing acute episodes. Prophylactic measures such as influenza vaccine are important in preventing respiratory infections and added stress on the heart. Other common treatment measures have been outlined earlier in this chapter. Maintaining an appropriate diet with a low sodium intake, low cholesterol, adequate protein and iron, and sufficient fluids is essential. Antianxiety drugs or sedatives may be useful. Depending on the underlying problem, cardiac support is provided by drugs previously mentioned. Medications such as ACE inhibitors can reduce renin secretion and vasoconstriction, digoxin improves cardiac efficiency, antihypertensives and vasodilators reduce blood pressure, and diuretics decrease sodium and water accumulations. Because patients often take a number of medications on a long-term basis, it is important to check all of them for effectiveness, cumulative toxicities, and interactions.

THINK ABOUT 12-9

a. Explain how cor pulmonale may develop.
b. Explain two causes of left-sided heart failure, one related to the heart and one systemic.
c. How should a patient with left-sided heart failure be positioned in a reclining chair or bed for treatment?

Congenital Heart Defects

Cardiac *anomalies* are structural defects in the heart that develop during the first 8 weeks of embryonic life. A structure such as a valve may be altered or missing. Several specific examples are described following this introduction. It is estimated that in the United States, 8 of every 1000 infants (approximately 40,000 babies) per year are born with heart defects, the majority of which are mild. Heart defects are the major cause of death in the first year of life. Mortality rates have dropped considerably with improvements in surgical procedures. Both genetic and environmental factors contribute to the occurrence of congenital heart defects and these defects often occur with other developmental problems.

Pathophysiology

Congenital heart disease may include valvular defects that interfere with the normal flow of blood (Fig. 12-23), septal defects that allow mixing of oxygenated blood from the pulmonary circulation with unoxygenated blood from the systemic circulation, shunts or abnormalities in position or shape of the large vessels (aorta and pulmonary artery), or combinations of these (Fig. 12-24). Selected examples follow. Most defects can be detected by the presence of heart murmurs. All significant defects result in a decreased oxygen supply to the tissues unless adequate compensations are available. If untreated, the child may develop heart failure.

Many variations and degrees of severity are possible with these defects, but if the basic cardiac cycle is understood, the effects of a change in blood flow in each situation can be predicted. Different methods of classifying the defects are possible, using either the type of defect

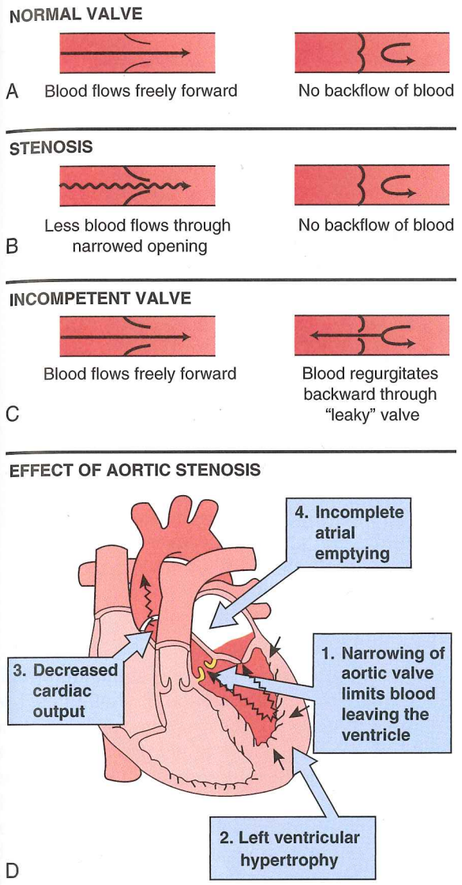

FIGURE 12-23 Effects of heart valve defects.

or the presence of cyanosis, a bluish color in the lips and oral mucosa.

When an abnormal communication permits mixing of blood, the fluid always flows from a high-pressure area to a low-pressure area, and flow occurs only in one direction. For example, a left-to-right shunt means that blood from the left side of the heart is recycled to the right side and to the lungs, resulting in an increased volume in the pulmonary circulation, a decreased cardiac output, and an inefficient system. On the other hand, a right-to-left shunt means that unoxygenated blood from the right side of the heart bypasses the lungs directly and enters the left side of the heart. The direction and amount of the abnormal blood flow determine the effects on the individual.

Acyanotic conditions are disorders in which systemic blood flow consists of oxygenated blood, although the amount may be reduced. In cyanotic disorders, venous blood mixes with arterial blood, permitting significant amounts of unoxygenated hemoglobin in the blood to bypass the lungs and enter the systemic circulation. The high proportion of unoxygenated blood produces a bluish color (characteristic of cyanosis) in the skin and mucous membranes, particularly the lips and nails. Death occurs in infancy in some severe cases, but many anomalies can be treated successfully shortly after birth.

Etiology

Most defects appear to be multifactorial and reflect a combination of genetic and environmental influences. These defects are often associated with chromosomal abnormalities, such as Down syndrome. Environmental factors include viral infections such as rubella, maternal alcoholism (fetal alcohol syndrome), and maternal diabetes.

Compensation Mechanisms

Through a sympathetic response, the heart increases its rate and force of contraction in an effort to increase cardiac output. This response increases the oxygen demand in the heart, restricts coronary perfusion, and increases peripheral resistance. The heart dilates and becomes hypertrophied. However, this response is ineffective because of the defect in the heart itself. Respiratory rate increases if the oxygen deficit results in acidosis due to increased lactic acid in the body, but oxygen levels must drop considerably before this factor influences the respiratory rate (see Chapter 13). Secondary polycythemia develops with chronic hypoxia as erythropoietin secretion increases as compensation.

Signs and Symptoms

Small defects are asymptomatic other than the presence of a heart murmur. Large defects lead to:

- Pallor and cyanosis
- Tachycardia, with a very rapid sleeping pulse and frequently a pulse deficit
- Dyspnea on exertion and tachypnea, in which the signs of heart failure are often present
- A squatting position, often seen in toddlers and older children, that appears to modify blood flow and be more comfortable for them
- Clubbed fingers (thick, bulbous fingertips) developed in time
- A marked intolerance for exercise and exposure to cold weather
- Delayed growth and development

Diagnostic Tests

Congenital defects, particularly severe ones, may be diagnosed at birth, but others may not be detected for some time. Many techniques and modalities, both invasive and noninvasive, can be used. Cardiomegaly can be observed on radiography. Examination techniques include diagnostic imaging, cardiac catheterization, echocardiograms, and ECG.

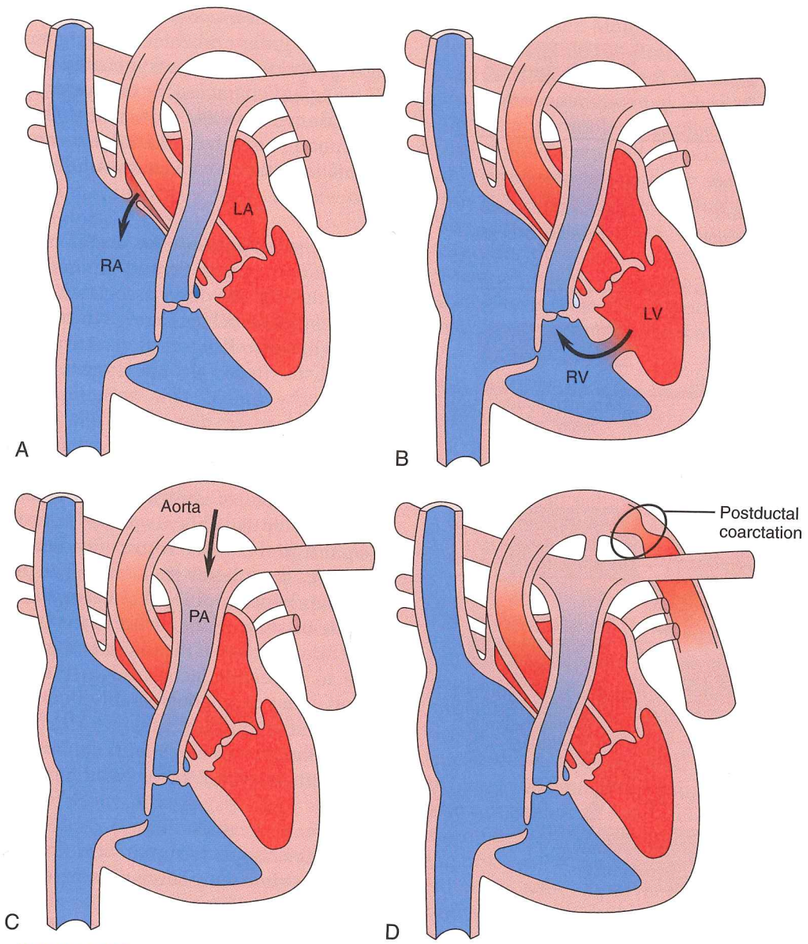

FIGURE 12-24 **A**, Atrial septal defect. Blood flow through the defect is usually left to right and produces an acyanotic shunt. **B**, Ventricular septal defect. Blood flow through the defect is usually left to right and produces an acyanotic shunt. **C**, Patent ductus arteriosus. Blood flow through the ductus is usually from the aorta to the pulmonary artery and produces an acyanotic shut. **D**, Coarction of the aorta. The arterial narrowing can produce a weaker pulse in lower extremities.

Treatment

Surgical repair is often needed to close abnormal openings or to replace valves or parts of vessels. Palliative surgery may take place immediately and then is followed up several years later by additional surgery. The timing of surgery depends on the individual situation, the severity of the defect, the ability of the individual to withstand surgery, and the impact of surgery on growth. In some cases, septal defects close spontaneously with time. Supportive measures and drug therapy are similar to those used for CHF. Prophylactic antimicrobial therapy may be administered before certain invasive procedures to prevent bacterial endocarditis (see Endocarditis in this chapter).

Ventricular Septal Defect

Ventricular septal defect (VSD) is the most common congenital heart defect and is commonly called a "hole in the heart." It is an opening in the interventricular septum, which may vary in size and location. (Septal defects may also occur in the atrial septum when the foramen ovale fails to close after birth.) Small defects do not affect cardiac function significantly but increase the risk of infective endocarditis.

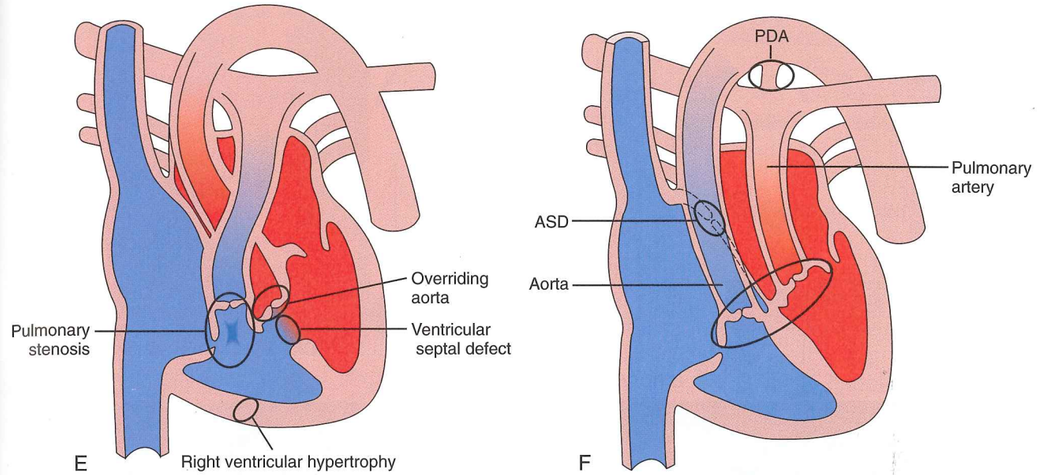

FIGURE 12-24, cont'd **E**, Tetralogy of Fallot showing the four characteristic abnormalities: pulmonary stenosis, ventricular septal defect, overriding aorta, and right ventricular hypertrophy. Tetralogy of Fallot is a cyanotic defect. **F**, Transposition of the great arteries. Two separate circulations are formed, which is incompatible with lilfe unless mixing blood occurs through other defects. *(From Copstead LE, Banisik J:* Pathophysiology, *ed 5, St. Louis, 2013, Saunders.)*

Large openings permit a *left-to-right* shunt of blood (see Fig. 12-24*A*). Blood can flow in *only one direction*, from the *high-pressure area* to the *low-pressure area*. In this case, the *left* ventricle is the high-pressure area, and therefore blood flows through the septal defect from the left ventricle to the right ventricle. The effect of this altered flow is that less blood leaves the left ventricle, reducing stroke volume and cardiac output to the systemic circulation. More blood enters the pulmonary circulation, some of which is already oxygenated; this reduces the efficiency of the system and in time overloads and irreversibly damages the pulmonary blood vessels, causing pulmonary hypertension. This complication, which may occur in untreated VSD, would lead to an abnormally high pressure in the right ventricle and a reversal of the shunt to a right-to-left shunt, leading to cyanosis.

THINK ABOUT 12-10

a. Describe the altered blood flow in the presence of an atrial septal defect. Include the direction of flow and the type of blood present in each circulation.
b. Patent ductus arteriosus results when the ductus arteriosus, a vessel between the aorta and the pulmonary artery that is present during fetal development, fails to close after birth. Using your knowledge of normal anatomy, trace the abnormal pattern of blood flow, including the rationale for it. Would a heart murmur be present?

Valvular Defects

Malformations most commonly affect the aortic and pulmonary valves. Valve problems may be classified as *stenosis*, or narrowing of a valve, which restricts the forward flow of blood, or valvular *incompetence*, which is a failure of a valve to close completely, allowing blood to regurgitate or leak backward (see Fig. 12-23). Mitral valve *prolapse* is a common occurrence; it refers to abnormally enlarged and floppy valve leaflets that balloon backward with pressure or to posterior displacement of the cusp, which permits regurgitation of blood. An effect similar to stenosis arises from abnormalities of the large vessels near the heart; for example, in coarctation (constriction) of the aorta.

Valvular defects reduce the efficiency of the heart "pump" and reduce stroke volume. If the opening is narrow, as in pulmonary stenosis, the myocardium must contract with more force to push the blood through (see Fig. 12-23 B and 12-24*B*). In time, that heart chamber will hypertrophy and may eventually fail. If a valve leaks and blood regurgitates backward, the heart must also increase its efforts to maintain cardiac output.

Mitral stenosis and its effects are demonstrated in an echocardiogram in Figure 12-25. Part *B* shows the normal valves and heart wall. In comparison, part *C* illustrates the thickened mitral valve leaflets and the narrow opening into the left ventricle. The left atrium is enlarged from the backup pressure and the increased workload has produced the thickened atrial wall.

Surgical replacement by mechanical, animal (porcine) or tissue engineered valves is often done (Fig. 12-26).

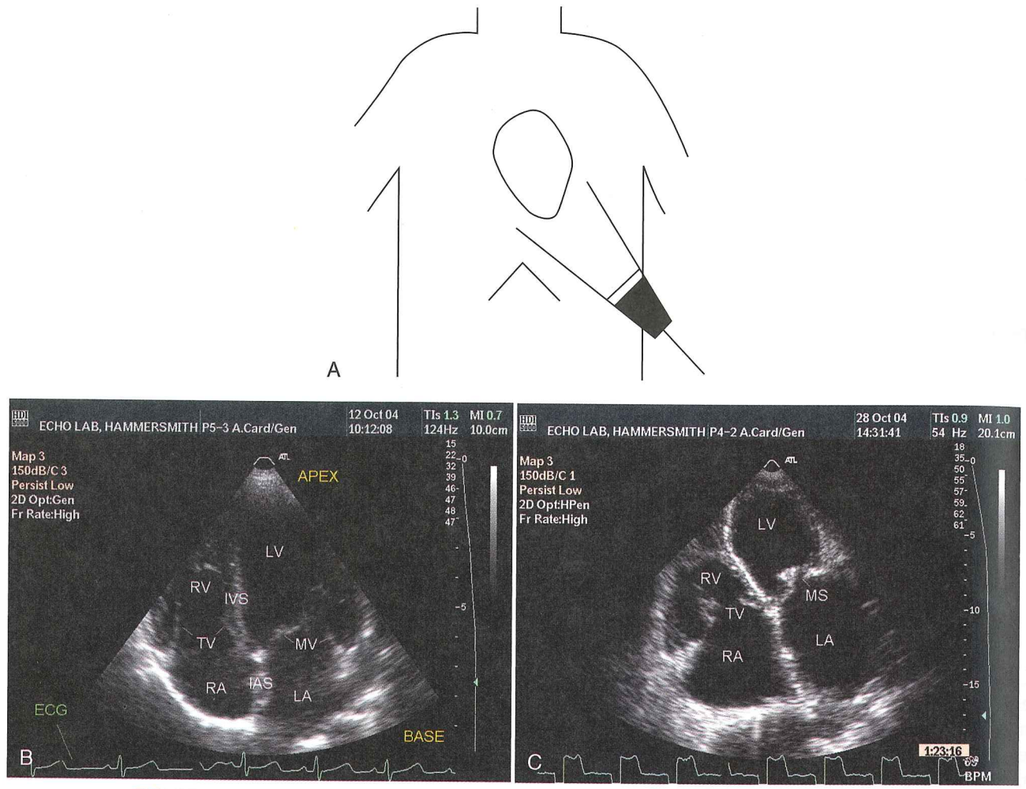

FIGURE 12-25 Echocardiograph showing mitral stenosis. *LA*, Left atrium; *LV*, left ventricle; *MS*, mitral stenosis; *MV*, mitral valve; *RA*, right atrium; *RV*, right ventricle; *TV*, tricuspid valve. **A**, Position of the transducer at the apical window. **B**, A 2-D image of a normal heart from the apical window, showing the four chambers and atrioventricular valves. An ECG is taken at the same time. **C**, The heart of a patient with mitral stenosis, indicated by thickening of the mitral valve leaflets, hypertrophy of the atrial wall, and enlargement of the atrial chambers. Note the change in the ECG indicating the cardiac phase affected by the abnormality. *(Courtesy of Helen Armstrong-Brown and Dr. P. Nihoyannopolous, Hammersmith Hospital, London, England.)*

These prosthetic valves may last up to 10 years, but are susceptible to thrombus formation, requiring patients to take daily ASA (see Fig. 12-26*B*). Also, infectious endocarditis is a risk, so prophylactic antimicrobial drugs are suggested before any procedure that might cause bacteremia.

THINK ABOUT 12-11

a. Explain why an incompetent valve reduces the efficiency of the heart contraction.
b. Would symptomatic mitral valve prolapse cause a cyanotic or an acyanotic condition? Explain your reasoning.

Tetralogy of Fallot

Tetralogy of Fallot is the most common cyanotic congenital heart condition. It is more complex and more serious than the others described so far because it includes four (Greek *tetra*) abnormalities and is a cyanotic disorder (infants are sometimes called "blue babies"). The four defects are pulmonary valve stenosis, VSD, dextroposition of the aorta (to the right over the VSD), and right ventricular hypertrophy (see Fig. 12-24*C*). This combination alters pressures within the heart and therefore alters blood flow.

The pulmonary valve stenosis restricts outflow from the right ventricle, leading to right ventricular hypertrophy and high pressure in the right ventricle. This pressure, now higher than the pressure in the left ventricle, leads to a right-to-left shunt of blood through the VSD. The flow of unoxygenated blood from the right ventricle directly into the systemic circulation is promoted by the position of the aorta, over the septum or VSD. The end result is that the pulmonary circulation

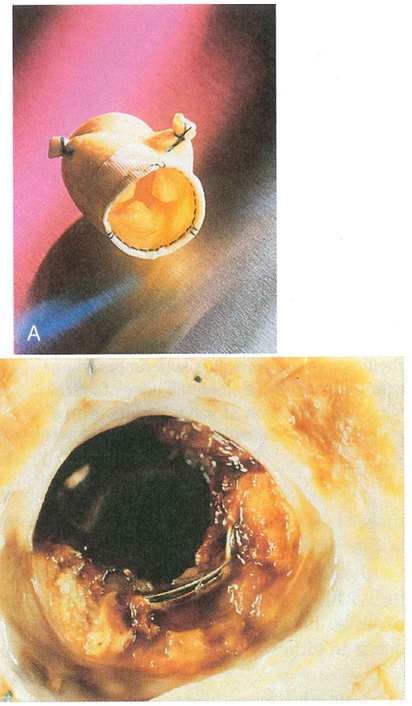

FIGURE 12-26 **A**, Porcine (pig) heart valve used to replace defective human valve. *(From deWit S, Kumagai C:* Medical-Surgical Nursing: Concepts and Practice, *Philadelphia, 2013, Saunders.)* **B**, Thrombosis obstructs mechanical prosthetic heart valve. *(From Kumar V, Abbas AK, Fausto M:* Robbins and Cotran Pathologic Basis of Disease, *ed 7, Philadelphia, 2005, Saunders.)*

receives a small amount of unoxygenated blood from the right ventricle, and the systemic circulation receives a larger amount of blood consisting of mixed oxygenated and unoxygenated blood. The oxygen deficit is great; hence, there are marked systemic effects and cyanosis.

THINK ABOUT 12-12

a. List the four defects present in the tetralogy of Fallot and state the effect each has on blood flow.
b. Describe the altered path of blood flow.
c. How does cyanosis occur with the altered blood flow?
d. Describe three signs of CHF in infants.

Inflammation and Infection in the Heart

Rheumatic Fever and Rheumatic Heart Disease

Pathophysiology

Rheumatic fever is an acute systemic inflammatory condition that appears to result from an abnormal immune

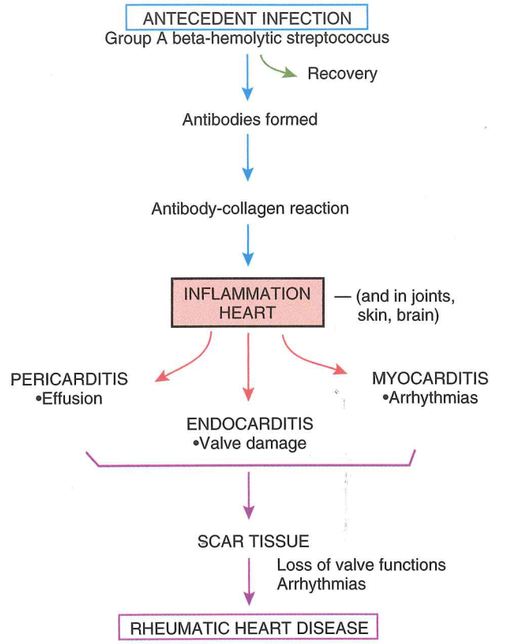

FIGURE 12-27 Development of rheumatic fever and rheumatic heart disease.

reaction occurring a few weeks after an untreated infection, usually caused by certain strains of group A beta-hemolytic *Streptococcus* (see Figs. 6-1*A* and 6-2*B*). The inflammation involves the heart and other parts such as joints and skin. It usually occurs in children 5 to 15 years of age. Although rheumatic fever occurs less frequently now in many areas, it remains a threat because new strains of *Streptococcus*, the cause of the antecedent infection, continue to appear. Also the long-term effects, seen as rheumatic heart disease, may be complicated by infective endocarditis and heart failure in older adults.

The antecedent or preceding infection commonly appears as an upper respiratory infection, tonsillitis, pharyngitis, or strep throat (awareness of the risk of rheumatic fever has led to increased use of rapid tests to quickly identify and treat a strep infection). Antibodies to the streptococcus organisms form as usual and then react with connective tissue (collagen) in the skin, joints, brain, and heart, causing inflammation (Fig. 12-27). The heart is the only site where scar tissue occurs, causing rheumatic heart disease.

During the acute stage, the inflammation in the heart may involve one or more layers of the heart:

- Pericarditis, inflammation of the outer layer, may include effusion (excessive fluid accumulation), which impairs filling.

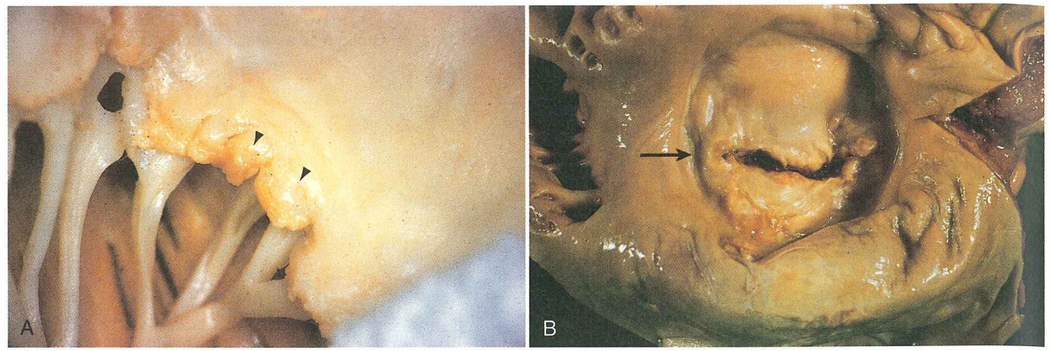

FIGURE 12-28 Rheumatic heart disease. **A**, Acute rheumatic mitral valvulitis superimposed on rheumatic heart disease (note thick cords). Verrucae *(arrows)* are visible along the edge of mitral valve leaflet. **B**, Mitral stenosis with fibrous thickening and distortion of valve leaflets. Arrow marks commissural fusion. *(From Kumar V, Abbas AK, Fausto M:* Robbins and Cotran Pathologic Basis of Disease, *ed 7, Philadelphia, 2005, Saunders.)*

- Myocarditis, in which the inflammation develops as localized lesions in the heart muscle, called Aschoff bodies, may interfere with conduction.
- Endocarditis, the most common problem, affects the valves, which become edematous, and *verrucae* form. Verrucae are rows of small, wartlike vegetations along the outer edge of the valve cusps (Fig. 12-28*A*). The mitral valve is affected most frequently. The inflammation disrupts the flow of blood and the effectiveness of the left ventricle. Eventually, the valve may be scarred, leading to stenosis if the cusps fuse together or to incompetence if fibrous tissue shrinks, or to a combination of these, ending in rheumatic heart disease (see Fig. 12-28*B*). In some cases the chordae tendineae are involved in the inflammatory reaction, and fibrosis ensues, leading to shortened chordae and malfunctioning valve. Recurrent inflammation is likely to cause more damage to the valves and increase risk for infective endocarditis.

Other sites of inflammation in patients with rheumatic fever include the:

- Large joints, particularly in the legs, which may be involved with synovitis in a migratory polyarthritis (often multiple joints affected)
- Skin, which may show a nonpruritic rash known as erythema marginatum (red macules or papules that enlarge and have white centers)
- Wrists, elbows, knees, or ankles, where small, nontender subcutaneous nodules usually form on the extensor surfaces
- Basal nuclei in the brain (more frequently in girls) causing involuntary jerky movements of the face, arms, and legs (Sydenham's chorea or Saint Vitus' dance)

Not all signs and symptoms occur in a single individual. Diagnosis is based on the presence of several of the preceding criteria including general signs of inflammatory disease, as well as high levels of antistreptolysin O antibodies and a history of prior streptococcal infection.

Rheumatic heart disease develops years later in some individuals, when scarred valves or arrhythmias compromise heart function. Congestive heart failure may occur in either the acute or chronic stage.

Signs and Symptoms

The general indications of a systemic inflammation are usually present in acute rheumatic fever:

- low-grade fever
- Leukocytosis
- Malaise
- Anorexia, and fatigue
- Tachycardia, even at rest, is common.
- Heart murmurs indicate the site of inflammation.
- Epistaxis and abdominal pain may be present. Acute heart failure may develop from the dysrhythmias or severe valve distortion. Recovery often requires a prolonged period of rest and treatment.

Diagnostic Tests

Elevated serum antibody levels remain after the infection has been eradicated (antistreptolysin O titer). Leukocytosis and anemia are common. Heart function tests, as previously mentioned, may be required. Characteristic ECG changes develop.

Treatment

Antibacterial agents such as penicillin V may be administered to eradicate any residual infection and prevent additional infection. Penicillin may be continued for some time to prevent recurrences, depending on previous attacks and the time lapse, the risk of infection, and the presence of cardiac damage. Any future streptococcal infection should be promptly treated. Anti-inflammatory agents such as ASA or corticosteroids (prednisone) may be given. Specific treatment is required for dysrhythmias or heart failure, as previously described.

Potential complications, such as heart failure resulting from severe valve damage, are similar to those mentioned earlier under congenital heart defects. Valve replacement may be necessary (see Fig. 12-26). The prognosis depends on the severity of heart damage and prevention of recurrences.

When valve damage has occurred, precautionary measures such as prophylactic penicillin prior to invasive procedures or dental treatment marked by significant bleeding are recommended to prevent bacteremia and infective endocarditis (see section on Infective Endocarditis).

THINK ABOUT 12-13

a. Describe the stages of development of acute rheumatic fever.
b. Explain how valvular damage due to rheumatic fever may be asymptomatic until increased exercise or pregnancy occurs.

Infective Endocarditis

Pathophysiology

Infective endocarditis, formerly called bacterial endocarditis, occurs in two forms: the subacute type, in which defective heart valves are invaded by organisms of low virulence, such as *Streptococcus viridans* (part of the normal flora of the mouth); and the acute type, in which normal heart valves are attacked by highly virulent organisms, such as *Staphylococcus aureus*, which tend to cause severe tissue damage and may be difficult to treat successfully. It is now recognized that many different types of organisms can cause infective endocarditis, and it is important to identify and treat the specific organism promptly.

The basic effects are the same, regardless of the causative organism. Microorganisms in the general circulation attach to the endocardium and invade the heart valves, causing inflammation and formation of vegetations on the cusps. Vegetations are large, fragile masses made up of fibrin strands, platelets and other blood cells, and microbes. In the acute stage, these may interfere with the opening and closing of the valves. Pieces may break away, forming infective or septic emboli that then cause infarction and infection in other tissues. This process causes additional destruction and scarring of the valve and the chordae tendineae.

Etiology

A combination of factors predisposes to infection: the presence of abnormal tissue in the heart, the presence of microbes in the blood, and reduced host defenses.

Abnormal valves associated with many predisposing conditions increase the risk of subacute infective endocarditis. These conditions include congenital defects, rheumatic fever, mitral prolapse, and artificial or replacement valves. Persons with septal defects, catheters, or other artificial implants are also susceptible toinfection.

Some individuals should be premedicated with penicillin or another antibacterial drug before any instrumentation or invasive procedure such as scaling of the teeth, in which a transient bacteremia could occur. Specific recommendations have been issued by the American Heart Association and American Dental Association regarding the conditions and procedures under which prophylactic medication should be given, the recommended drugs, and dosing. Medical conditions are classified with regard to the degree of risk for endocarditis. The drug of choice for prophylaxis is amoxicillin taken 1 hour before the procedure; the alternative in cases of allergy to penicillin is clindamycin or cephalexin.

Abscesses or other sources of infection should be treated promptly. Intravenous drug users have an increased incidence of acute endocarditis. Anyone in whom the immune system is suppressed, such as those taking corticosteroids or those with acquired immunodeficiency syndrome, is vulnerable. Endocarditis, both bacterial and fungal, is also a risk with open heart surgery.

THINK ABOUT 12-14

Explain why a tooth extraction or scaling procedure could pose a special danger to an individual with an altered heart valve.

Signs and Symptoms

Various new heart murmurs are the common indicator, as well as other signs of impaired heart function. Initially it may be difficult to detect any change in the heart murmur from the predisposing condition, but the increasing impairment soon affects the sounds. Transesophageal echocardiogram may also be used to reveal the presence of vegetations. Subacute infective endocarditis is frequently insidious in onset, manifesting only as an intermittent low-grade fever or fatigue. Anorexia, splenomegaly, and Osler's nodes (painful red nodules on the fingers) are often present. Septic emboli from the vegetations that cause vascular occlusion or infection and abscesses in other areas of the body, will result in additional manifestations depending on the location of the secondary problem. The release of bacteria into the blood may lead to intermittent increased fever. Congestive heart failure develops in severe cases.

Acute endocarditis has a sudden, marked onset, with spiking fever, chills, and drowsiness. Heart valves are badly damaged causing severe impairment of heart function. As in the subacute form, septic emboli may cause infarctions or abscesses in organs, resulting in appropriate signs related to location.