



FIGURE 11-8 (A) Atrial fibrillation illustrating absence of distinct P waves together with irregular ventricular rate, usually 140 to 160 beats per minute. (B) Ventricular fibrillation illustrating extremely abnormal chaotic cardiac rhythm without any evidence of synchronized electrical impulses.
Garcia, T. B., and Holtz, N. E. 2003. *Introduction to 12-Lead ECG*. Boston: Jones and Bartlett Publishers.

Heart Disease: An Introduction

Heart disease can result from congenital (and sometimes inherited) defects in the structure of the heart or in the contractile apparatus of cardiac myocytes. Infectious agents may damage heart valves and myocytes. Atherosclerotic disease of the coronary arteries will lead to compromised blood supply to the heart, termed coronary ischemia. If severe, such ischemia will result in cardiac myocyte necrosis, an event that defines a myocardial infarct (heart attack) with sequelae that may result in sudden cardiac death or some degree of chronic heart disease and failure. Systemic hypertension (excessive systemic blood pressure) will cause the heart to compensate by undergoing left ventricular hypertrophy. Ultimately the heart may not be able to compensate for the extra workload and fails.

Congenital Heart Disease

CARDIAC DEVELOPMENT AND PRENATAL BLOOD FLOW

The heart undergoes a complex developmental sequence. It is formed from a tube that undergoes segmental dilations and constrictions along with considerable growth and change in configuration. Eventually the individual chambers, valves, and large arteries develop, culminating in the final structural characteristics of a normal fully developed heart.

As the heart is developing, the blood flow through the fetal heart differs from its final postdelivery flow pattern. Much of the blood flow in the pulmonary artery

Ductus arteriosus A fetal artery connecting the pulmonary artery with the aorta that permits pressure determined blood flow from pulmonary artery into the aorta, bypassing blood flow to the nonfunctional fetal lungs.

Foramen ovale An opening in the atrial septum covered by a one-way flap valve regulated by pressure differences between the atria, permitting blood flow from right to left atrium but not in the opposite direction, thereby bypassing blood flow from right cardiac chambers to the nonfunctional fetal lungs.

is diverted away from the lungs, which are nonfunctional in the fetus, and used instead to supply other fetal tissues. One bypass, called the **ductus arteriosus**, is a large communication connecting the pulmonary artery with the aorta that shunts much of the blood pumped into the pulmonary artery directly into the aorta. As soon as the infant is born and begins to breathe air, the lungs expand and the ductus arteriosus constricts, blocking blood flow. Consequently, pulmonary artery blood can then flow only into the newly expanded lungs; the nonfunctional ductus eventually becomes converted into a fibrous cord called the ligamentum arteriosum. The other bypass is an opening in the atrial septum called the **foramen ovale**, which maintains blood flow between the two atria as the atrial septum is developing. Blood (in part derived from maternal circulation via the umbilical vein and hence oxygen-rich) flow across the foramen ovale is controlled by a flap of atrial tissue on the left atrial side of the septum that covers (overlaps) the opening in the foramen ovale. In this position, the flap functions as a one-way valve that allows blood to flow from the right atrium into the left atrium but does not allow flow in the opposite direction (FIGURE 11-9). The right-to-left flow is determined by pressure differences between the two chambers. In the fetus, the blood pressure is higher in the right atrium than in the left atrium because only a relatively small volume of blood flows through the lungs and is returned to the left atrium. Most is directed into the aorta through the ductus arteriosus. After birth, the left atrial pressure rises when the lungs expand, allowing a large volume of blood to flow through the lungs and into the left atrium.

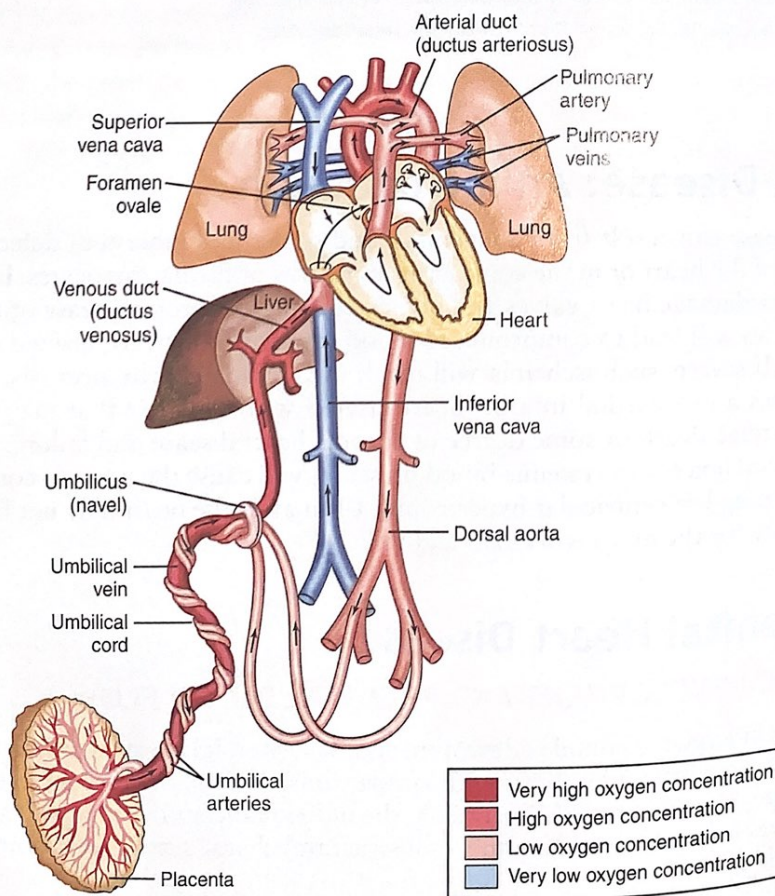


FIGURE 11-9 General pattern of fetal circulation.

The higher left atrial pressure presses the flap valve against the left atrial surface of the septum, closing the communication between the atria. Usually, the tissue flap fuses with the atrial septum to form a solid partition between the two atria. Sometimes, the fusion is incomplete but no flow of blood from right to left atria is possible as long as the left atrial pressure exceeds the pressure in the right atrium, which holds the flap against the atrial septum.

PATHOGENESIS AND MANIFESTATIONS OF CONGENITAL HEART DISEASE

Sometimes the heart fails to develop normally. Partitions between cardiac chambers may be defective, the cardiac valves may be malformed, or the large vessels entering and leaving the heart may not communicate normally with the appropriate atrium or ventricle. Some viral infections, such as rubella (German measles) or other maternal illnesses during the early phases of fetal development, may cause improper development of the heart as well as other organs. Some drugs or medications taken by the mother may disrupt normal fetal development. Some chromosomal abnormalities, such as Down syndrome, also are frequently associated with abnormal cardiac development. Genetic factors may account for some cardiac abnormalities, but often the reason for a congenital abnormality cannot be determined.

The effect of a structural abnormality depends on the nature of the defect and its effect on the circulation of blood. Most people with congenital heart abnormalities have a heart murmur caused by turbulent flow of blood within the heart, related to the cardiac malformation. Many congenital heart abnormalities result from abnormal communications between the systemic and pulmonary circulations that permit blood to be shunted between the adjacent chambers. The amount of blood shunted and the direction of the shunt depend on the size of the opening between the chambers, and the blood pressure difference between the chambers determines the direction of flow.

Most shunts are left-to-right shunts from left cardiac chambers (systemic circulation) into right cardiac chambers (pulmonary circulation). A left-to-right shunt mixes oxygenated blood from the left cardiac chambers with deoxygenated blood in the right chambers, but the admixture does not affect the oxygen content of the blood delivered to the tissues by the left ventricle. The amount of blood shunted depends on the size of the septal defect. A small defect shunts very little blood and has no significant effect on cardiovascular function. However, a large septal defect can shunt a large volume of blood, which puts an additional burden on the right ventricle, which is overfilled by the shunted blood. The larger volume of blood pumped into the lungs raises the pulmonary blood pressure, which eventually damages the lungs by causing thickening and narrowing of the pulmonary blood vessels. As the pulmonary vascular damage progresses and the pulmonary artery pressure continues to rise, the right ventricle has to work even harder to overcome the increasing resistance to blood flow through the lungs. The higher right ventricular pressure also causes the right atrial pressure to rise, reducing the pressure differences between the left and right atria. Consequently, the amount of blood shunted from the left to the right atrium also falls. Little or no blood shunting occurs when left and right atrial pressures equalize, and if right atrial pressure exceeds left atrial pressure, blood shunts between the atria in the opposite direction resulting in deoxygenated blood entering the circulation (a right-to-left shunt). This is termed **Eisenmenger syndrome** and has serious consequences.

Right-to-left shunts mix poorly oxygenated blood from the right cardiac chambers with normally oxygenated blood contained in the left cardiac chambers, which reduces the oxygen content of the blood pumped by the left ventricle to supply the

Eisenmenger syndrome
Blood shunts between the atria in the opposite direction of normal.

body. The affected person's activities usually are severely restricted because of the low oxygen content of the arterial blood. The skin and mucous membranes acquire a blue color called **cyanosis**, which is caused by the low oxygen saturation of arterial blood, and congenital cardiovascular abnormalities associated with cyanosis are grouped together under the general term cyanotic congenital heart disease.

COMMON CARDIOVASCULAR ABNORMALITIES

The more common and important cardiovascular abnormalities fall into four major groups: failure of the normal fetal bypass channels to close; defects in the formation of the atrial and ventral septae; abnormalities that obstruct blood flow through the heart, pulmonary artery, or aorta; and malformations of the aorta and pulmonary artery or abnormal connection of the arteries to the appropriate ventricles.

Patent Ductus Arteriosus

Normally the ductus closes spontaneously soon after birth in full-term infants, in part due to falling levels of prostaglandins. A large patent ductus shunts blood from the aorta into the pulmonary artery and causes the same clinical manifestations and complications as an intracardiac left-to-right shunt. This is treated by the use of prostaglandin synthesis inhibitors (such as indomethacin) or, if necessary, surgical closure of the ductus.

Atrial and Ventricular Septal Defects

Usually, an atrial septal defect results from defective development of the partitions that divide the atria with the defect located in the middle of the septum at the site usually occupied by the foramen ovale. Small defects in children often close spontaneously and are often asymptomatic as discussed above. Larger defects should be closed, which usually can be accomplished using a device inserted into the heart through a peripheral vein. Sometimes an open surgical procedure is required to place a patch over the defect.

A common type of atrial septal defect is a patent foramen ovale. The foramen ovale normally becomes nonfunctional after birth, caused by the rapid postdelivery changes in atrial pressures. The left atrial pressure rises when pulmonary blood flow increases after the ductus arteriosus closes. The higher left atrial pressure pushes the flap valve of the foramen ovale against the atrial septum, where it usually fuses with the septum. In newborn infants, the foramen ovale may remain patent and functional if the infant has a congenital cardiac abnormality that is associated with a high right atrial pressure, which forces right-to-left blood flow through the foramen ovale. In about 25 percent of adults, the flap valve does not fuse completely, but the foramen ovale remains nonfunctional as long as the left atrial pressure remains higher than the right atrial pressure.

Ventricular septal defects are also very common (FIGURE 11-10). Many are less than 3 mm in diameter and often close spontaneously, but larger defects require surgical closure. Generally, all large septal defects need to be closed because of the harmful effects of a large defect.

Pulmonary or Aortic Valve Stenosis Caused by Semilunar Valve Maldevelopment

Abnormal development of the semilunar valve leaflets narrows the valve opening, which can vary from 2 to 10 mm diameter. The degree of obstruction depends on the diameter of the orifice (FIGURE 11-11). Pulmonary stenosis obstructs outflow from the right ventricle, and aortic stenosis impedes outflow from the left ventricle. Treatment consists of dilating the valve opening by inserting a balloonlike device into the narrow opening.

Valvular Heart Disease

Rheumatic valvular heart disease has markedly declined, along with rheumatic fever, but other conditions that cause valve malfunction have assumed greater importance. These include various degenerative conditions of the aortic valve and an abnormality of the mitral valve that causes it to prolapse into the atrium during ventricular systole.

RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE

Rheumatic fever is a complication of infection by the group A beta hemolytic streptococcus, the organism responsible for streptococcal sore throat and scarlet fever. Clinically, the affected individual has an acute arthritis affecting multiple joints (which is why the disease is called “rheumatic” fever) and evidence of inflammation of the heart.

Rheumatic fever is not a bacterial infection but a type of hypersensitivity reaction induced by various antigens present in the streptococcus. This reaction develops several weeks after the initial streptococcal infection. Apparently, some individuals form antibodies against antigens present in the streptococcus, and the antistreptococcal antibody cross-reacts with similar antigens in the individual’s own tissues. The antigen–antibody reaction injures connective tissue and is responsible for the febrile illness. Fortunately, rheumatic fever develops in only a small proportion of people with group A beta streptococcal infections.

Patients with acute rheumatic fever rarely die as a result of severe inflammation of the heart and consequent acute heart failure; generally the fever and signs of inflammation eventually subside although healing is often associated with some degree of scarring. In the joints and in many other tissues, scarring causes no difficulties, but scarring of heart valves may produce deformities that impair function.

Rheumatic heart disease is caused by scarring of the heart valves subsequent to the healing of a rheumatic inflammation. This complication is relatively common and primarily affects the valves of the left side of the heart, the mitral and aortic valves (FIGURE 11-16). If the valve does not close properly, blood refluxes back through it (called **regurgitation**). Frequently, the damaged valve also does not open properly, and the valve orifice is narrowed. This is called a valve **stenosis**. Valve lesions impair cardiac function. When valvular stenosis is present, the heart must exert more effort than normal to force blood through the narrowed orifice. In regurgitation, a portion of the ventricular output is not expelled normally and leaks through the incompetent valve. This is a serious disadvantage because the heart must repump the volume of regurgitated blood to deliver the same amount of blood to the peripheral tissues.

When a person is seriously disabled by a rheumatic valvular deformity, it is possible to excise the abnormal, scarred heart valve surgically and replace it with an artificial valve (discussed later in this chapter).

NONRHEUMATIC AORTIC STENOSIS

Nonrheumatic aortic stenosis can be caused by aortic stenosis secondary to a bicuspid aortic valve or by the aging of the valve. In about 2 percent of people, the aortic valve has two rather than the usual three cusps. This abnormality is called a congenital bicuspid aortic valve. The valve functions satisfactorily for a time but is subjected to unusual stress during opening and closing because of its bicuspid configuration. As a result, the valve gradually becomes thickened and may eventually become calcified after many years, leading to marked rigidity of the valve when a person

Rheumatic
A disease caused by hypersensitivity of the beta chain of the streptococcus, characterized by joint pains, and inflammation of the heart valve.

Regurgitation
Backflow of blood through a valve that does not close properly.

Stenosis
Narrowing of a valve orifice.

coarctation is much higher than normal because the heart has to pump blood at a much higher pressure to deliver blood through the narrowed segment of aorta, possibly leading to left ventricular disease. The pressure and volume of blood flowing into the aorta distal to the coarctation are both lower than normal, and a collateral circulation develops to bypass the obstruction. An individual with a coarctation may appear normal except for high blood pressure identified when measuring pressure in the brachial arteries, but lower-than-normal blood pressure in the arteries of the lower extremities. Usually, the narrowed segment of aorta is relatively short and can be treated by resecting the constricting segment and reconnecting the aorta so that its caliber is normal throughout its entire length.

The Tetralogy of Fallot and Transposition of the Great Arteries

Both of these conditions result from abnormal division of a single channel called the truncus arteriosus, which extends from the developing ventricles and will be divided by a partition to form the aorta and the pulmonary artery. The partition takes a spiral course as it divides the truncus arteriosus, which is why the aorta and the pulmonary artery spiral around each other as they attach to their respective ventricles. The two abnormalities caused by abnormal division of the truncus arteriosus are relatively common, and both cause intermixing of deoxygenated blood with oxygenated blood, which leads to marked cyanosis and related problems.

The tetralogy of Fallot results if the septum that divides the truncus is misplaced and divides unequally. This results in the four abnormalities that comprise the tetralogy: a ventricular septal defect, pulmonary stenosis, an enlarged aorta that overrides the septal defect, and right ventricular hypertrophy that develops as a consequence of the pulmonary stenosis. In this condition, poorly oxygenated blood in the right ventricle flows through the septal defect to mix with blood from the left ventricle flowing into the aorta, which overrides the septal defect. Treatment consists of enlarging the opening of the narrowed pulmonary artery and closing the septal defect.

Transposition of the great arteries results if the septum that divides the truncus arteriosus does not follow its normal spiral course when it divides the truncus into the aorta and pulmonary artery. Consequently, the aorta and pulmonary artery develop parallel to each other. The aorta becomes located to the right of the pulmonary artery instead of behind and to the left of the pulmonary artery. The aorta becomes connected to the right ventricle and the pulmonary artery attaches to the left ventricle, which severely disrupts blood flow in both the pulmonary and systemic circulations. Consequently, the body is supplied by poorly oxygenated blood that is continuously circulated in the systemic circulation and the flow of oxygenated blood remains confined to the pulmonary circuit, where it serves no useful purpose. After birth, the condition is not compatible with life unless there is a communication that permits some intermixing of blood between the pulmonary and systemic circulations such as a patent foramen ovale, atrial septal defect, or ventricular septal defect (FIGURE 11-12). Generally, such communications do not provide enough oxygenated blood to supply the infant's needs.

The current treatment of this condition is called the arterial switch operation in which the aorta is connected to the left ventricle, and the pulmonary artery is attached to the right ventricle. It is also necessary to reposition the coronary arteries so they are connected properly to the artery supplying blood to the left ventricle.

TABLE 11-1 summarizes the principal features of the congenital cardiovascular malformations described in this section, which are also illustrated in FIGURE 11-13.