PROFESSIONAL GUIDE TO **DISEASES**

ELEVENTH EDITION

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CLINICAL EDITOR

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To my teachers and mentors, past and present,
and to those patients who have truly touched
my life with a difficult diagnosis or a tough
conversation—this work is for you.

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FOREWORD

How can health care providers respond to the new model of consumerism in medicine, in which patients are encouraged to question everything, are bombarded with health care information in the media, and are targeted by direct advertising campaigns marketing everything from weight loss programs to therapies for erectile dysfunction? In today's information- and technology-driven environment, staying on the leading edge of ever-changing medical information can be overwhelming. The reality of contemporary medical economics and regulation of medical practice is driving many clinicians to see greater numbers of patients each day, each with more complex illnesses. This has resulted in less time spent with the patient, less time to perform detailed literature searches for complex conditions, and more time devoted to medical record completion, negotiating with medical insurance companies and other administrative representatives and justifying the medical necessity for many orders. In this environment there's a need for a well-organized and comprehensive summary of a wide variety of illnesses to help the busy clinician.

This 11th edition of *Professional Guide to Diseases* features well-organized chapters coordinated around disease clusters. The data in this edition are accurate, updated in terms of original research and practice guidelines, and designed to provide a brief yet comprehensive overview of a large array of disease processes and new sections for a quick pathophysiology review. Readers ranging from the trainee preparing for a board certification examination to the senior faculty member or other health care provider needing a ready reference will find that this book provides a clear and concise overview of hundreds of diseases and conditions.

This edition features improved sections focused on health promotion, disease prevention, and special focuses on elderly patients. Additionally, there is an updated section on bioterrorism and a new section on approaches to care of LGBTQ individuals—topics that have been receiving added emphasis in health care circles in recent years. Complications associated with different diseases are also included. This edition also updates information on many conditions for which a variety of clinical treatment guidelines have been published recently by major professional medical and surgical organizations. The content of clinical practice guidelines typically includes a detailed review of the literature for the specific topic.

For the variety of health care providers for whom this book has been created, the organizational format allows the option of reviewing an entire topic, such as clinical genetics and genomics, or narrowing the focus to one aspect of disease, such as the details of treatment or special considerations of emerging illnesses. The text is written clearly and includes graphic data displays that make it useful as a quick reference, but it also contains comprehensive material that allows for more complete coverage of a group of similar disease entities. In this edition of the book are artwork and figures that illustrate the pathophysiology of many conditions in a full-color format. These images can not only be used to better understand the topic on a professional level, but can also be leveraged for patient

teaching while in the office or hospital. This book continues to feature information on efficient health care delivery for routine conditions seen almost daily, as well as cultural considerations in patient care, information on potential bioterrorism agents, updates on rare diseases, and a revised section discussing complementary and alternative therapies for specific conditions.

As the practice of medicine continues to evolve, providers must face the challenges of achieving correct diagnoses using more targeted and cost-effective testing in a more constricted time frame. The 11th edition of *Professional Guide to Diseases* offers a strong background of successful editions combined with the latest updates and recommendations that will help you provide accurate information to your patients.

Enjoy this book!

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Cardiovascular Disorders

Introduction

The cardiovascular system begins its activity when the fetus is barely a month old and is the last body system to cease activity at the end of life. This system is so vital that its activity defines the presence of life.

Life-Giving Transport System

The heart, arteries, veins, and lymphatics form the cardiovascular network that serves as the body's transport system, bringing life-supporting oxygen and nutrients to cells, removing metabolic waste products, and carrying hormones from one part of the body to another. Often called the *circulatory system*, it may be divided into two branches: *pulmonary circulation*, in which blood picks up new oxygen and liberates the waste product carbon dioxide; and *systemic circulation* (including coronary circulation), in which blood carries oxygen and nutrients to all active cells while transporting waste products to the kidneys, liver, and skin for excretion.

Circulation requires normal functioning of the heart, which propels blood through the system by continuous rhythmic contractions. Located behind the sternum, the heart is a muscular organ the size of a man's fist. It has three layers: the *endocardium*—the smooth inner layer; the *myocardium*—the thick, muscular middle layer that contracts in rhythmic beats; and the *epicardium*—the thin, serous membrane, or outer surface of the heart. Covering the entire heart is a saclike membrane called the *pericardium*, which has two layers: a *visceral* layer that's in contact with the heart and a *parietal*, or outer, layer. To prevent irritation when the heart moves against this layer during contraction, fluid lubricates the parietal pericardium.

The heart has four chambers: two thin-walled chambers called *atria* and two thick-walled chambers called *ventricles*. The atria serve as reservoirs during ventricular contraction (systole) and as booster pumps during ventricular relaxation (diastole). The left ventricle propels blood through the systemic circulation. The right ventricle, which forces blood through the pulmonary circulation, is much thinner than the left because it meets only one sixth the resistance.

ELDER TIP *As a person's body ages, the ventricular and aortic walls stiffen, decreasing the heart's pumping action.*

Heart Valves

Two kinds of valves work inside the heart: *atrioventricular* (AV) and *semilunar*. The AV valve between the right atrium and the ventricle has three leaflets, or cusps, and three papillary muscles; hence, it's called the *tricuspid valve*. The AV valve between the left atrium and the ventricle consists of two cusps shaped like a bishop's hat, or miter, and two papillary muscles and is called the *mitral valve*. The tricuspid and mitral valves prevent blood backflow from the ventricles to the atria during ventricular contraction. The leaflets of both valves are attached to the ventricles' papillary muscles by thin, fibrous bands called *chordae tendineae*; the leaflets separate and descend funnel-like into the ventricles during diastole and are pushed upward and together during systole to occlude the mitral

and tricuspid orifices. The valves' action isn't entirely passive because papillary muscles contract during systole and prevent the leaflets from prolapsing into the atria during ventricular contraction.

The two semilunar valves, which resemble half-moons, prevent blood backflow from the aorta and pulmonary arteries into the ventricles when those chambers relax and fill with blood from the atria. They're referred to as the *aortic valve* and *pulmonic valve* for their respective arteries.

ELDER TIP *In elderly people, fibrotic and sclerotic changes thicken heart valves and reduce their flexibility. These changes lead to rigidity and incomplete closure of the valves, which may result in systolic or diastolic murmurs.*

THE CARDIAC CYCLE

Diastole is the phase of ventricular relaxation and filling. As diastole begins, ventricular pressure falls below arterial pressure, and the aortic and pulmonic valves close. As ventricular pressure continues to fall below atrial pressure, the mitral and tricuspid valves open, and blood flows rapidly into the ventricles. Atrial contraction then increases the volume of ventricular filling by pumping 15% to 25% more blood into the ventricles. When *systole* begins, the ventricular muscle contracts, raising ventricular pressure above atrial pressure and closing the mitral and tricuspid valves. When ventricular pressure finally becomes greater than that in the aorta and pulmonary artery, the aortic and pulmonic valves open, and the ventricles eject blood. Ventricular pressure continues to rise as blood is expelled from the heart. As systole ends, the ventricles relax and stop ejecting blood, and ventricular pressure falls, closing both valves.

S_1 (the first heart sound) is heard as the ventricles contract and the AV valves close. S_1 is loudest at the heart's apex, over the mitral area. S_2 (the second heart sound), which is normally rapid and sharp, occurs when the aortic and pulmonic valves close. S_2 is loudest at the heart's base (second intercostal space [ICS] on both sides of the sternum).

Ventricular distention during diastole, which can occur in systolic heart failure, creates low-frequency vibrations that may be heard as a third heart sound (S_3), or ventricular gallop. An atrial gallop (S_4) may appear at the end of diastole, just before S_1, if atrial filling is forced into a ventricle that has become less compliant or overdistended or has a decreased ability to contract. A pressure rise and ventricular vibrations cause this sound.

CARDIAC CONDUCTION

The heart's conduction system is composed of specialized cells capable of generating and conducting rhythmic electrical impulses to stimulate heart contraction. This system includes the sinoatrial (SA) node, the AV junction, the bundle of His and its bundle branches, and the ventricular conduction tissue and Purkinje fibers.

Normally, the SA node controls the heart rate and rhythm at 60 to 100 beats/minute. Because the SA node has the lowest resting potential, it's the heart's pacemaker. If it defaults, another part of the system takes over. The AV junction may emerge at 40 to 60 beats/minute; the bundle of His and bundle branches at 30 to 40 beats/minute; and ventricular conduction tissue at 20 to 30 beats/minute.

ELDER TIP *As the myocardium of the aging heart becomes more irritable, extrasystoles may occur along with sinus arrhythmias and sinus bradycardias. In addition, increased fibrous tissue infiltrates the SA nodes and internodal atrial tracts, which may cause atrial fibrillation and flutter.*

CARDIAC OUTPUT

Cardiac output—the amount of blood pumped by the left ventricle into the aorta each minute—is calculated by multiplying the *stroke volume* (the amount of blood the left ventricle ejects during each contraction) by the *heart rate* (number of beats/minute). When cellular demands increase, stroke volume or heart rate must increase.

Many factors affect the heart rate, including exercise, pregnancy, and stress. When the sympathetic nervous system releases norepinephrine, the heart rate increases; when the parasympathetic system releases acetylcholine, it slows. As a person ages, the heart rate takes longer to normalize after exercise.

Stroke volume depends on the ventricular blood volume and pressure at the end of diastole (preload), resistance to ejection (afterload), and the myocardium's contractile strength (inotropy). Changes in preload, afterload, or inotropic state can alter the stroke volume.

ELDER TIP *Exercise cardiac output declines slightly with age. A decrease in maximum heart rate and contractility may cause this change.*

CIRCULATION AND PULSES

Blood circulates through three types of vessels: *arteries, veins*, and *capillaries.* The sturdy, pliable walls of the arteries adjust to the volume of blood leaving the heart. The major artery branching out of the left ventricle is the aorta.

Its segments and subbranches ultimately divide into minute, thin-walled (one-cell thick) capillaries. Capillaries pass the blood to the veins, which return it to the heart. In the veins, valves prevent blood backflow.

ELDER TIP *Aging contributes to arterial and venous insufficiency as the strength and elasticity of blood vessels decrease.*

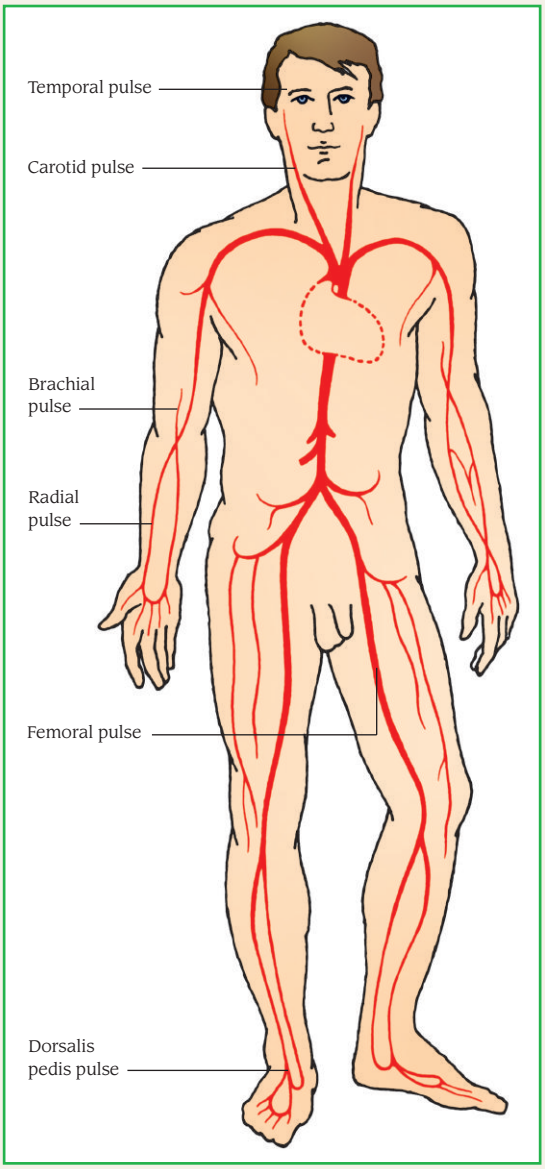
Pulses are felt best wherever an artery runs near the skin and over a hard structure. (See *Pulse points*.) Easily found pulses are:

- *radial artery*—anterolateral aspect of the wrist
- *temporal artery*—in front of the ear, above and lateral to the eye
- *common carotid artery*—neck (side)
- *femoral artery*—groin

Pulse points

Peripheral pulse rhythm should correspond exactly to the auscultatory heart rhythm. The pulse's character may offer useful information. For example, *pulsus alternans*, a strong beat followed by a weak one, can mean myocardial weakness. A *water-hammer* (or *Corrigan*) *pulse*, a forceful bounding pulse best felt in the carotid arteries or in the forearm, accompanies increased pulse pressure—commonly with capillary pulsations of the fingernails (*Quincke sign*). This pulse usually indicates aortic valve regurgitation.

Pulsus bisferiens, a double peripheral pulse for every apical beat, can signal aortic regurgitation or hypertrophic obstructive cardiomyopathy. *Pulsus bigeminus* is a coupled rhythm; you feel its beat in pairs. *Pulsus paradoxus* is exaggerated waxing and waning of the arterial pressure (≥15 mm Hg decrease in systolic blood pressure during inspiration), often seen in cardiac tamponade.



The lymphatic system also plays a role in the cardiovascular network. Originating in tissue spaces, the lymphatic system drains fluid and other plasma components that build up in extravascular spaces and reroutes them back to the circulatory system as lymph, a plasmalike fluid. Lymphatics also extract bacteria and foreign bodies.

CARDIOVASCULAR ASSESSMENT

Physical assessment provides vital information about cardiovascular status.

◆ Check for underlying cardiovascular disorders, such as central cyanosis (impaired gas exchange), edema (heart failure or valvular disease), and clubbing (congenital cardiovascular disease).

◆ Palpate the peripheral pulses bilaterally and evaluate their rate, equality, and quality on a scale of 0 (absent) to +4 (bounding). (See *Pulse amplitude scale*.)

◆ Inspect the carotid arteries for equal appearance. Auscultate for bruits; then palpate the arteries individually, one side at a time, for *thrills* (fine vibrations due to irregular blood flow).

◆ Check for pulsations in the jugular veins (more easily seen than felt). Watch for jugular vein distention (JVD)—a possible sign of right-sided heart failure, valvular stenosis, cardiac tamponade, or pulmonary embolism. Take blood pressure readings in both arms while the patient is lying, sitting, and standing.

◆ Palpate the precordium for any abnormal pulsations, such as lifts, heaves, or thrills. Use the palms (at the base of the fingertips) or the fingertips. The normal apex will be felt as a light tap and extends over 1″ (2.5 cm) or less.

◆ Systematically auscultate the anterior chest wall for each of the four heart sounds in the aortic area (second ICS at the right sternal border), pulmonic area (second ICS at the left sternal border), right ventricular area (lower half of the left sternal border), and mitral area (fifth ICS at the midclavicular line). However, don't limit your auscultation to these four areas. Valvular sounds may be heard all over the precordium. Therefore, inch your stethoscope in a Z pattern, from the base of the heart across and down and then over to the apex, or start at the apex and work your way up. For low-pitched sounds, use the bell of the stethoscope; for high-pitched sounds, the diaphragm. Carefully inspect each area for pulsations, and palpate for thrills. Check the location of apical pulsation for deviations in normal size ($^3/_8$″ to $^3/_4$″ [1 to 2 cm]) and position (in the mitral area)—possible signs of left ventricular hypertrophy, left-sided valvular disease, or right ventricular disease.

Pulse amplitude scale

To record your patient's pulse amplitude, use this standard scale:

0: Pulse isn't palpable.
+**1:** Pulse is thready, weak, difficult to find, may fade in and out, and disappears easily with pressure.
+**2:** Pulse is constant but not strong; light pressure must be applied or pulse will disappear.
+**3:** Pulse considered normal. Is easily palpable, doesn't disappear with pressure.
+**4:** Pulse is strong, bounding, and doesn't disappear with pressure.

◆ Listen for the vibrating sound of turbulent blood flow through a stenotic or incompetent valve. Time the murmur to determine where it occurs in the cardiac cycle—between S_1 and S_2 (systolic), between S_2 and the following S_1 (diastolic), or throughout systole (holosystolic). Finally, listen for the scratching or squeaking of a pericardial friction rub.

SPECIAL CARDIOVASCULAR TESTS

Electrocardiography (ECG) measures electrical activity by recording currents transmitted by the heart. It can detect ischemia, injury, necrosis, bundle branch blocks, fascicular blocks, conduction delay, chamber enlargement, and arrhythmias. In Holter monitoring, a tape recording tracks as many as 100,000 cardiac cycles over a 12- or 24-hour period. This test may be used to assess the effectiveness of antiarrhythmic drugs or to evaluate arrhythmia symptoms. A signal-averaged ECG will identify afterpotentials, which are associated with a risk of ventricular arrhythmias. (See *Positioning chest electrodes*, page 5.)

Chest X-rays may reveal cardiac enlargement and aortic dilation. They also assess pulmonary circulation. When pulmonary venous and arterial pressures rise, characteristic changes appear, such as dilation of the pulmonary venous shadows. When pulmonary venous pressure exceeds oncotic pressure of the blood, capillary fluid leaks into lung tissues, causing pulmonary edema. This fluid may settle in the alveoli, producing a butterfly pattern, or the lungs may appear cloudy or hazy; in the interlobular septa, sharp linear densities (Kerley lines) may appear.

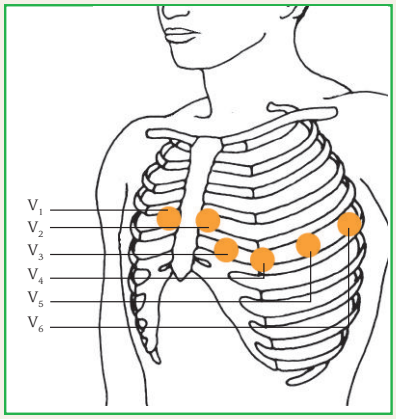
Exercise testing intra-aortic balloon pump (IABP) treadmill determines the heart's response to physical stress. This test measures blood pressure and ECG changes during

Positioning chest electrodes

To record the 12-lead electrocardiogram, place electrodes on the patient's arms and legs (with the ground lead on the patient's right leg). The three standard limb leads (I, II, III) and the three augmented leads (aV_R, aV_L, aV_F) are recorded using these electrodes.

To record the precordial (chest) leads, place the electrodes as follows:

- V_1—fourth ICS, right sternal border
- V_2—fourth ICS, left sternal border
- V_3—midway between V_2 and V_4
- V_4—fifth ICS, left midclavicular line
- V_5—fifth ICS, left anterior axillary line
- V_6—fifth ICS, left midaxillary line



increasingly rigorous exercises. Myocardial ischemia, abnormal blood pressure response, or arrhythmias indicate the circulatory system's failure to adapt to exercise.

Cardiac catheterization evaluates chest pain, the need for coronary artery surgery or angioplasty, congenital heart defects, and valvular heart disease and determines the extent of heart failure. Right-sided catheterization involves threading a pulmonary artery thermodilution catheter, which can measure cardiac output, through a vein into the right side of the heart, pulmonary artery, and its branches in the lungs to measure right atrial, right ventricular, pulmonary artery, and pulmonary artery wedge pressures (PAWPs). Left-sided catheterization entails retrograde catheterization of the left ventricle or transseptal catheterization of the left atrium. Ventriculography during left-sided catheterization involves injecting radiopaque dye into the left ventricle to measure ejection fraction and to disclose abnormal heart wall motion or mitral valve incompetence.

In coronary angiography, radiopaque material injected into coronary arteries allows cineangiographic visualization of coronary arterial narrowing or occlusion.

Echocardiography uses echoes from pulsed high-frequency sound waves (ultrasound) to evaluate cardiac structures. Two-dimensional echocardiography (most common), in which an ultrasound beam rapidly sweeps through an arc, produces a cross-sectional or fan-shaped view of cardiac structures. Contrast agents may be used for image enhancement. Doppler echocardiography records blood flow within the cardiovascular system. Color Doppler echocardiography shows the direction of blood flow, which provides information about the degree of valvular insufficiency. Transesophageal echocardiography combines ultrasound with endoscopy to better view the heart's structures. This procedure allows images to be taken from the heart's posterior aspect.

Echocardiography provides information about valve leaflets, size and dimensions of heart chambers, and thickness and motion of the septum and the ventricular walls. It can also reveal intracardiac masses, detect pericardial effusion, diagnose hypertrophic cardiomyopathy, and estimate cardiac output and ejection fraction. This test can also evaluate possible aortic dissection when it involves the ascending aorta.

In multiple-gated acquisition scanning (MUGA), a radioactive isotope in the intravascular compartment allows measurement of stroke volume, wall motion, and ventricular ejection fraction. Myocardial imaging uses a radioactive isotope to detect abnormalities in myocardial perfusion. This agent concentrates in normally perfused areas of the myocardium but not in ischemic areas ("cold spots"), which may be permanent (scar tissue) or temporary (from transient ischemia). These tests can be done as exercise studies or can be combined with drugs (nuclear stress test), in patients unable to exercise.

Peripheral arteriography consists of a fluoroscopic X-ray after arterial injection of a contrast

medium. Similarly, phlebography defines the venous system after injection of a contrast medium into a vein.

Doppler ultrasonography evaluates the peripheral vascular system and assesses peripheral artery disease (PAD) when combined with sequential systolic blood pressure readings.

Endomyocardial biopsy can detect cardiomyopathy, infiltrative myocardial diseases, and, most often, transplant rejection.

Electrophysiologic studies help diagnose conduction system disease and serious arrhythmias. Electronic induction and termination of arrhythmias aid drug selection. Endocardial mapping detects an arrhythmia's focus using a finger electrode. Epicardial mapping uses a computer and a device containing electrodes that's slipped over the heart to detect arrhythmias.

Magnetic resonance imaging (MRI) can investigate cardiac structure and function. Positron emission tomography and magnetic resonance spectroscopy are used to assess myocardial metabolism.

Electron-beam computed tomography, also known as ultrafast computed tomography, is used to detect microcalcifications in the coronary arteries and can give a coronary calcium score. This test is useful for identifying early coronary artery disease (CAD).

Blood tests

Cardiac enzymes (cellular proteins released into blood after cell membrane injury) confirm acute myocardial infarction (MI) or severe cardiac trauma. All cardiac enzymes—creatine kinase (CK), lactate dehydrogenase, and aspartate aminotransferase, for example—are also found in other cells. Fractionation of enzymes can determine the source of damaged cells. For example, three fractions of CK are isolated, one of which (an isoenzyme called *CK-MB*) is found only in cardiac cells. CK-MB in the blood indicates injury to myocardial cells.

Measurement of a cardiac protein called *troponin* is the most precise way to determine whether a patient has experienced an MI. Some 6 hours after an MI, a blood test can detect two forms of troponin: T and I. Troponin T levels peak about 2 days after an MI and return to normal about 16 days later. Troponin I levels reach their peak in less than 1 day after an MI and return to normal in about 7 days.

Managing cardiovascular disease

Patients with cardiovascular disease pose a tremendous challenge. Their sheer numbers alone compel a thorough understanding of cardiovascular anatomy, physiology, and pathophysiology. Anticipate a high anxiety level in cardiac patients, and provide support and reassurance, especially during procedures such as cardiac catheterization.

Cardiac rehabilitation programs are widely prescribed and offer education and support along with exercise instruction. Rehabilitation programs begin in healthcare facilities and continue on an outpatient basis. Helping the patient resume a satisfying lifestyle requires planning and comprehensive teaching. Inform the patient about healthcare facilities and organizations that offer cardiac rehabilitation programs.

Congenital acyanotic defects

Ventricular septal defect

Causes and incidence

In ventricular septal defect (VSD), the most common congenital heart disorder, an opening in the septum between the ventricles allows blood to shunt between the left and the right ventricles. This disease accounts for up to 30% of all congenital heart defects. Although most children with congenital heart defects are otherwise normal, in some, VSD coexists with additional birth defects, especially Down syndrome and other autosomal trisomies, renal anomalies, and such cardiac defects as patent ductus arteriosus (PDA) and coarctation of the aorta. VSDs are located in the membranous or muscular portion of the ventricular septum and vary in size. Some defects close spontaneously; in other defects, the entire septum is absent, creating a single ventricle. The prognosis is good for defects that close spontaneously or are correctable surgically but poor for untreated defects, which are sometimes fatal by age 1, usually from secondary complications. Less than 1% of neonates are born with VSD. In 80% to 90% of neonates who are born with this disorder, the hole is small and will usually close spontaneously. In the remaining 10% to 20% of neonates, surgery is needed to close the hole. (See *Understanding ventricular septal defect*, page 7.)

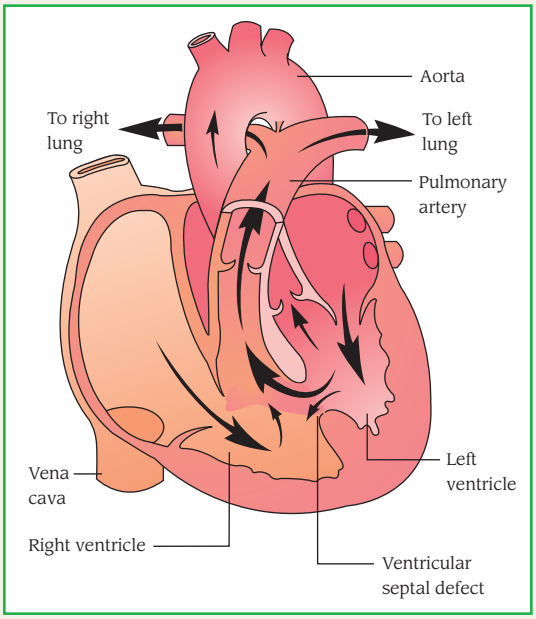
Pathophysiology

In neonates with VSD, the ventricular septum fails to close completely by the eighth week of gestation, as it would normally.

VSD isn't readily apparent at birth, because right and left ventricular pressures are about equal, so blood doesn't shunt through the defect. As the pulmonary vasculature gradually relaxes, 4 to 8 weeks after birth, right ventricular

Understanding ventricular septal defect

A ventricular septal defect (VSD), the most common type of congenital disorder, is an abnormal opening between the right and left ventricles that allows blood to shunt between them. Not always readily apparent at birth, the defect can be small and may close spontaneously. The septum may be entirely absent, resulting in a single ventricle. A large, untreated defect can cause right ventricular hypertrophy, pulmonary hypertension, and heart failure. VSD is classified as an increased pulmonary blood flow defect.



pressure decreases, allowing blood to shunt from the left to the right ventricle. In small or restrictive defects, right ventricle pressure is only slightly elevated, while pulmonary artery pressures (PAPs) and peripheral vascular resistance (PVR) remain normal. In moderate defects, the size of the defect determines the magnitude of the shunt. When right ventricular pressure decreases, the left atrium and ventricle may become fluid overloaded. Because large defects do not meet a lot of flow resistance, the pressure of the ventricles is equal at first. As PVR decreases, volume increases to the pulmonary system, in turn increasing the volume of the left ventricle. This, in turn, can create left ventricular dilation, followed by increased left atrial pressure and pulmonary venous pressure.

Complications

- Right arterial and ventricular hypertrophy
- Heart failure
- Pulmonary hypertension

Signs and symptoms

Clinical features of VSD vary with the defect's size, the shunting's effect on the pulmonary vasculature, and the infant's age. In a small VSD, shunting is minimal, and PAP and heart size remain normal. Such defects may eventually close spontaneously without ever causing symptoms.

Initially, large VSD shunts cause left atrial and left ventricular hypertrophy. Later, an uncorrected VSD will cause right ventricular hypertrophy due to increasing pulmonary vascular resistance. Eventually, biventricular heart failure and cyanosis (from reversal of shunt direction) occur. Resulting cardiac hypertrophy may make the anterior chest wall prominent. A large VSD increases the risk of pneumonia.

Infants with large VSDs are thin and small and gain weight slowly. They may develop heart failure with dusky skin; liver, heart, and spleen enlargement because of systemic venous congestion; diaphoresis; feeding difficulties; rapid, grunting respirations; and increased heart rate. They may also develop severe pulmonary hypertension. Fixed pulmonary hypertension may occur much later in life with right-to-left shunt (Eisenmenger syndrome), causing cyanosis and clubbing of the nail beds.

The typical murmur associated with a VSD is blowing or rumbling and varies in frequency. In the neonate, a moderately loud early systolic murmur may be heard along the lower left sternal border. About the second or third day

after birth, the murmur may become louder and longer. In infants, the murmur may be loudest near the heart's base and may suggest pulmonary stenosis (PS). A small VSD may produce a functional murmur or a characteristic loud, harsh systolic murmur. Larger VSDs produce audible murmurs (at least a grade 3 pansystolic), loudest at the fourth ICS, usually with a thrill; however, a large VSD with minimal pressure gradient may have no audible murmur. In addition, the pulmonic component of S_2 sounds loud and is widely split. Palpation reveals displacement of the point of maximal impulse to the left. When fixed pulmonary hypertension is present, a diastolic murmur may be audible on auscultation, the systolic murmur becomes quieter, and S_2 is greatly accentuated.

Diagnosis

Diagnostic findings include:

- Chest X-ray is normal in small defects; in large VSDs, it shows cardiomegaly, left atrial and left ventricular enlargement, and prominent pulmonary vascular markings.
- ECG is normal in children with small VSDs; in large VSDs, it shows left and right ventricular hypertrophy, suggesting pulmonary hypertension.
- Echocardiography may detect a large VSD and its location in the septum, estimate the size of a left-to-right shunt, suggest pulmonary hypertension, and identify associated lesions and complications.

CONFIRMING DIAGNOSIS *Cardiac catheterization determines the VSD's size and exact location, calculates the degree of shunting by comparing the blood oxygen saturation in each ventricle, determines the extent of pulmonary hypertension, and detects associated defects.*

Treatment

In mild cases, no treatment is needed, although the infant should be closely followed to make sure that the hole closes properly as the infant grows. Large defects usually require early surgical correction before heart failure and irreversible pulmonary vascular disease develop.

For small defects, surgery consists of simple suture closure. Moderate to large defects require insertion of a patch graft, using cardiopulmonary bypass. In patients with heart failure, digoxin and diuretics may be prescribed to control symptoms. In patients who develop increased pulmonary resistance and irreversible pulmonary vascular changes that produce a reversible right-to-left shunt (Eisenmenger syndrome), a heart–lung transplant may be required.

If the child has other defects and will benefit from delaying surgery, pulmonary artery banding normalizes pressures and flow distal to the band and prevents pulmonary vascular disease, allowing postponement of surgery. (Pulmonary artery banding is done only when the child has other complications.) A rare complication of VSD repair is complete heart block from interference with the bundle of His during surgery. (Heart block may require temporary or permanent pacemaker implantation.)

Before surgery, treatment consists of:

- digoxin, sodium restriction, and diuretics to prevent heart failure
- careful monitoring by physical examination, X-ray, and ECG to detect increased pulmonary hypertension, which indicates a need for early surgery
- measures to prevent infection (prophylactic antibiotics, e.g., to prevent infective endocarditis)

Generally, postoperative treatment includes a brief period of mechanical ventilation. The patient will need analgesics and may also require diuretics to increase urine output, continuous infusions of nitroprusside or adrenergic agents to regulate blood pressure and cardiac output and, in rare cases, a temporary pacemaker.

Special considerations

Although the parents of an infant with VSD often suspect something is wrong with their child before diagnosis, they need psychological support to help them accept the reality of a serious cardiac disorder. Because surgery may take place months after diagnosis, parent teaching is vital to prevent complications until the child is scheduled for surgery or the defect closes. Thorough explanations of all tests are also essential.

- Instruct parents to watch for signs of heart failure, such as poor feeding, sweating, and heavy breathing.
- If the child is receiving digoxin or other medications, tell the parents how to give it and how to recognize adverse effects. Caution them to keep medications out of the reach of all children.
- Teach parents to recognize and report early signs of infection and to avoid exposing the child to people with obvious infections.
- Encourage parents to let the child engage in normal activities.
- Tell parents to follow up with their pediatrician. Also tell them that child life therapy may be appropriate if their child displays delayed growth and development or failure to thrive.
- Stress the importance of prophylactic antibiotics before and after surgery.

After surgery to correct VSD:

- Monitor vital signs and intake and output. Maintain the infant's body temperature with an overbed warmer. Give catecholamines, nitroprusside, and diuretics, as ordered; analgesics as needed.
- Monitor central venous pressure (CVP), intra-arterial blood pressure, and left atrial or PAP readings. Assess heart rate and rhythm for signs of conduction block.
- Check oxygenation, particularly in a child who requires mechanical ventilation. Suction to maintain a patent airway and to prevent atelectasis and pneumonia, as needed.
- Monitor pacemaker effectiveness if needed. Watch for signs of failure, such as bradycardia and hypotension.
- Reassure parents and allow them to participate in their child's care.

ATRIAL SEPTAL DEFECT

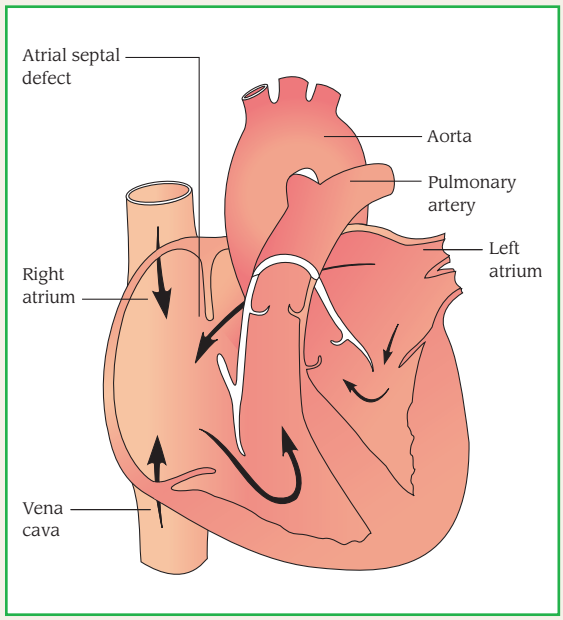
Causes and incidence

In an atrial septal defect (ASD), an opening between the left and the right atria allows shunting of blood between the chambers. *Ostium secundum defect* (most common) occurs in the region of the fossa ovalis and occasionally extends inferiorly, close to the vena cava; *sinus venosus defect* occurs in the superior–posterior portion of the atrial septum, sometimes extending into the vena cava, and is almost always associated with abnormal drainage of pulmonary veins into the right atrium; *ostium primum defect* occurs in the inferior portion of the septum primum and is usually associated with AV valve abnormalities (cleft mitral valve) and conduction defects. The cause of ASD is unknown.

ASD accounts for about 6% to 8% of congenital heart defects and appears almost twice as often in females as in males, with a strong familial tendency. Although ASD is usually a benign defect during infancy and childhood, delayed development of symptoms and complications makes it one of the most common congenital heart defects diagnosed in adults. ASD is present in 4 of every 100,000 people. Symptoms usually develop before age 30. When no other congenital defect exists, the patient—especially if a child—may be asymptomatic. The prognosis is excellent in asymptomatic patients but poor in those with cyanosis caused by large, untreated defects. (See *Understanding atrial septal defect.*)

Understanding atrial septal defect

An atrial septal defect (ASD) is an abnormal opening between the left and the right atria. A small opening may cause few symptoms. However, if the opening is large, higher pressure in the left atrium can shunt large amounts of blood into the right atrium, which can result in right heart volume overload, right atrial and ventricular enlargement, and pulmonary hypertension. ASD is classified as an increased pulmonary blood flow defect and is one of the most common congenital heart defects.



Pathophysiology

In this condition, blood shunts from left to right because left atrial pressure normally is slightly higher than right atrial pressure; this pressure difference forces large amounts of blood through a defect. The left-to-right shunt results in right heart volume overload, affecting the right atrium, right ventricle, and pulmonary arteries. Eventually, the right atrium enlarges, and the right ventricle dilates to accommodate the increased blood volume. If pulmonary artery hypertension develops because of the shunt (rare in children), increased pulmonary vascular resistance and right ventricular hypertrophy will follow. In some adult patients, irreversible (fixed) pulmonary artery hypertension causes reversal of the shunt direction, which results in unoxygenated blood entering the systemic circulation, causing cyanosis.

Complications

- Unoxygenated blood in systemic circulation
- Right and left ventricular hypertrophy
- Atrial arrhythmias
- Heart failure
- Emboli

Signs and symptoms

ASD commonly goes undetected in preschoolers; such children may complain about feeling tired only after extreme exertion and may have frequent respiratory tract infections but otherwise appear normal and healthy. However, children with large shunts may show growth retardation. Children with ASD seldom develop heart failure, pulmonary hypertension, infective endocarditis, or other complications. However, as adults, they usually manifest pronounced symptoms, such as fatigability and dyspnea on exertion, frequently to the point of severe limitation of activity (especially after age 40).

In children, auscultation reveals an early to midsystolic murmur, superficial in quality, heard at the second or third left ICS. In patients with large shunts (resulting from increased tricuspid valve flow), a low-pitched diastolic murmur is heard at the lower left sternal border, which becomes more pronounced on inspiration. Although the murmur's intensity is a rough indicator of the size of the left-to-right shunt, its low pitch sometimes makes it difficult to hear and, if the pressure gradient is relatively low, a murmur may not be detectable. Other signs include a fixed, widely split S_2, caused by delayed closure of the pulmonic valve, and a systolic click or late systolic murmur at the apex, resulting from mitral valve prolapse (MVP), which occasionally affects older children with ASD.

In older patients with large, uncorrected defects and fixed pulmonary artery hypertension, auscultation reveals an accentuated S_2. A pulmonary ejection click and an audible S_4 may also be present. Clubbing and cyanosis become evident; syncope and hemoptysis may occur with severe pulmonary vascular disease.

Diagnosis

A history of increasing fatigue and characteristic physical features suggest ASD. The following findings confirm it:

- Chest X-ray shows an enlarged right atrium and right ventricle, a prominent pulmonary artery, and increased pulmonary vascular markings.
- ECG may be normal but usually shows right axis deviation, prolonged PR interval, varying degrees of right bundle branch block, right ventricular hypertrophy, atrial fibrillation (particularly in severe cases after age 30) and, in ostium primum defect, left axis deviation.
- Echocardiography measures right ventricular enlargement, may locate the defect, and shows volume overload in the right side of the heart. (Other causes of right ventricular enlargement must be ruled out.)

CONFIRMING DIAGNOSIS *Two-dimensional echocardiography with color Doppler flow, contrast echocardiography, or both have supplanted cardiac catheterization as the confirming tests for ASD. Cardiac catheterization is used if inconsistencies exist in the clinical data or if significant pulmonary hypertension is suspected.*

Treatment

Operative repair is advised for all patients with uncomplicated ASD with evidence of significant left-to-right shunting. Ideally, this is performed when the patient is between ages 2 and 4. Operative treatment shouldn't be performed in patients with small defects and trivial left-to-right shunts. Because ASD seldom produces complications in infants and toddlers, surgery can be delayed until they reach preschool or early school age. A large defect may need immediate surgical closure with sutures or a patch graft.

Physicians have developed a new procedure, referred to as catheter closure or transcatheter closure of the ASD, that uses wires or catheters to close ASD without surgery. In this procedure, the surgeon makes a tiny incision in the groin to introduce the catheters, then advances the catheters into the heart, and places the closure device across the ASD. This procedure may not be applicable to all patients.

Special considerations

- Before cardiac catheterization, explain pretest and posttest procedures to the child and her parents. If possible, use drawings or other visual aids to explain it to the child.
- As needed, teach the patient about prophylactic antibiotics to prevent infective endocarditis. (They may be administered before dental or other invasive procedures.)
- If surgery is scheduled, teach the child and his or her parents about the intensive care unit (ICU) and introduce them to the staff. Show parents where they can wait during the operation. Explain postoperative procedures, tubes, dressings, and monitoring equipment.
- After surgery, closely monitor the patient's vital signs, central venous and intra-arterial pressures, and intake and output. Watch for atrial arrhythmias, which may remain uncorrected.

COARCTATION OF THE AORTA

Causes and incidence

Coarctation is a narrowing of the aorta, usually just below the left subclavian artery, near the site where the ligamentum arteriosum (the remnant of the ductus arteriosus, a fetal blood vessel) joins the pulmonary artery to the aorta. Coarctation may occur with aortic valve stenosis (usually of a bicuspid aortic valve) and with severe cases of hypoplasia of the aortic arch, PDA, and VSD. This is typically sporadic and without clear cause before this condition induces severe systemic hypertension or degenerative changes in the aorta. (See *Understanding coarctation of the aorta*.)

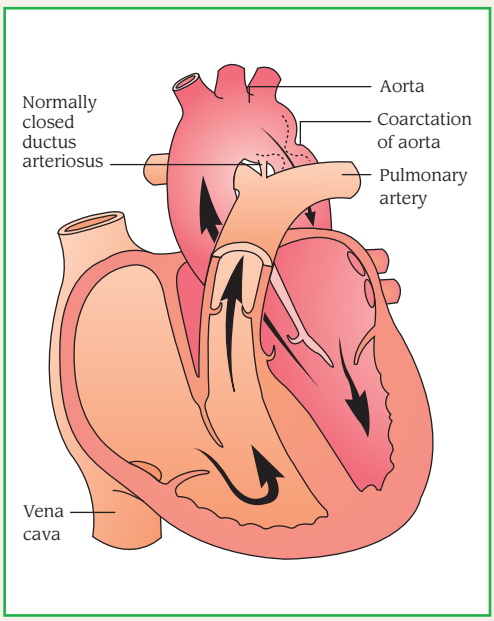
Coarctation of the aorta occurs in 4 of every 10,000 people born each year in the United States and is usually diagnosed in children or adults younger than age 40. It accounts for about 4% to 6% of all congenital heart defects in children and is twice as common in males as in females. When it occurs in females, it's commonly associated with Turner syndrome, a chromosomal disorder that causes ovarian dysgenesis. Generally, the prognosis for coarctation of the aorta depends on the severity of associated cardiac anomalies; the prognosis for isolated coarctation is good if corrective surgery is performed.

Pathophysiology

Coarctation of the aorta may develop as a result of spasm and constriction of the smooth muscle in the ductus arteriosus as it closes. Possibly, this contractile tissue extends into the aortic wall, causing narrowing. The obstructive process causes hypertension in the aortic branches

Understanding coarctation of the aorta

Coarctation is a narrowing of the aorta, usually just below the left subclavian artery, near the site where the ligamentum arteriosum joins the pulmonary artery to the aorta. It can result from spasm and constriction of the smooth muscle in the ductus arteriosus as it closes. Restricted blood flow through the narrow aorta increases the pressure load on the left ventricle, resulting in dilation of the proximal aorta, left ventricular hypertrophy, elevated upper body blood pressures, and diminished blood flow to the lower body. The ductus arteriosus may be open or closed. Coarctation of the aorta is more common in boys and is the leading cause of heart failure in the first few months of life. It's classified as an obstruction to blood flow leaving the heart.



above the constriction (arteries that supply the arms, neck, and head) and diminished pressure in the vessels below the constriction.

Restricted blood flow through the narrowed aorta increases the pressure load on the left ventricle and causes dilation of the proximal aorta and ventricular hypertrophy. Untreated, this condition may lead to left-sided heart failure and, rarely, to cerebral hemorrhage and aortic rupture. If VSD accompanies coarctation, blood shunts left to right, straining the right side of the heart. This leads to pulmonary hypertension and, eventually, right-sided heart hypertrophy and failure.

Complications

- Infective endocarditis
- Pulmonary hypertension
- Right ventricular hypertrophy
- Right-sided heart failure

Signs and symptoms

Clinical features vary with age. During the first year of life, when aortic coarctation may cause heart failure, the infant displays tachypnea, dyspnea, pulmonary edema, pallor, tachycardia, failure to thrive, cardiomegaly, and hepatomegaly. In most cases, heart sounds are normal unless a coexisting cardiac defect is present. Femoral pulses are absent or diminished.

If coarctation is asymptomatic in infancy, it usually remains so throughout adolescence, as collateral circulation develops to bypass the narrowed segment. During adolescence, this defect may produce dyspnea, claudication, headaches, epistaxis, and hypertension in the upper extremities despite collateral circulation. It commonly causes resting systolic hypertension and wide pulse pressure; high diastolic pressure readings are the same in both the arms and the legs. Coarctation may also produce a visible aortic pulsation in the suprasternal notch, a continuous systolic murmur, an accentuated S_2, and an S_4.

Diagnosis

CONFIRMING DIAGNOSIS *The cardinal signs of coarctation of the aorta are resting systolic hypertension, absent or diminished femoral pulses, and wide pulse pressure.*

The following tests support this diagnosis:

- Chest X-ray may demonstrate left ventricular hypertrophy, heart failure, a wide ascending and descending aorta, and notching of the undersurfaces of the ribs, due to extensive collateral circulation.
- ECG may eventually reveal left ventricular hypertrophy.
- Echocardiography may show increased left ventricular muscle thickness, coexisting aortic valve abnormalities, and the coarctation site.
- Doppler ultrasound and cardiac catheterization evaluate collateral circulation and measure pressure in the right and left ventricles and in the ascending and descending aortas (on both sides of the obstruction).
- MRI enables assessment of the anatomy and function of aortic abnormalities.

Treatment

For an infant with heart failure caused by coarctation of the aorta, treatment consists of medical management with digoxin, diuretics, oxygen, and sedatives. If medical management fails, surgery may be needed.

The child's condition usually determines the timing of surgery. Signs of heart failure or hypertension may call for early surgery. If these signs don't appear, surgery usually occurs during the preschool years.

Before the operation, the child may require endocarditis prophylaxis or, if he or she is older and has previously undetected coarctation, antihypertensive therapy. During surgery, the surgeon uses a flap of the left subclavian artery to reconstruct an unobstructed aorta.

Balloon angioplasty with possible stent placement may also be indicated for some patients as an alternative to surgical repair. It uses a technique similar to that used to open the coronary arteries, but is performed on the aorta.

Special considerations

- Palpate the pulses in the legs in newborns and at well-baby visits to detect absent or diminished pulses.
- When coarctation in an infant requires rapid digitalization, monitor vital signs closely and watch for digoxin toxicity (poor feeding and vomiting).
- Balance intake and output carefully, especially if the infant is receiving diuretics with fluid restriction.
- Because the infant may not be able to maintain proper body temperature, regulate environmental temperature with an overbed warmer if needed.
- Monitor blood glucose levels to detect possible hypoglycemia, which may occur as glycogen stores become depleted.
- Offer the parents emotional support and an explanation of the disorder. Also explain diagnostic procedures, surgery, and drug therapy. Tell parents what to expect postoperatively.
- For an older child, assess the blood pressure in extremities regularly, explain any exercise

restrictions, stress the need to take medications properly and to watch for adverse effects, and teach about tests and other procedures.

After corrective surgery:

- Monitor blood pressure closely, using an intra-arterial line. Measure blood pressure in arms and legs. Monitor intake and output.
- If the patient develops hypertension and requires a medication such as nitroprusside, administer it, as ordered, using an infusion pump. Watch for severe hypotension and regulate the dosage carefully.
- Provide pain relief and encourage a gradual increase in activity.
- Promote adequate respiratory functioning through turning, coughing, and deep breathing.
- Watch for abdominal pain or rigidity and signs of gastrointestinal (GI) or urinary bleeding.
- If an older child needs to continue antihypertensives after surgery, teach the patient and his parents about them.
- Stress the importance of continued endocarditis prophylaxis as appropriate.

PATENT DUCTUS ARTERIOSUS

Causes and incidence

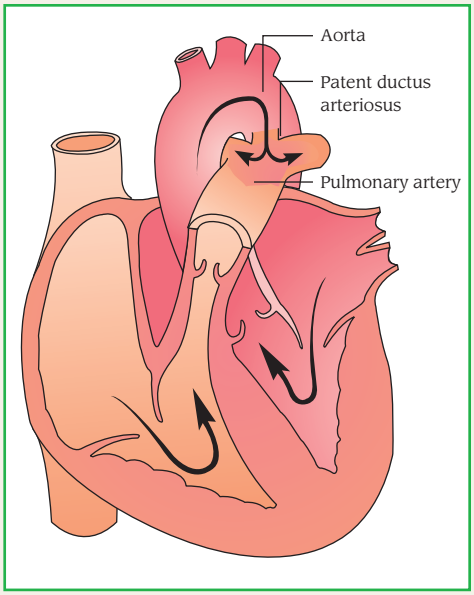
The ductus arteriosus is a fetal blood vessel that connects the pulmonary artery to the descending aorta. In PDA, the lumen of the ductus remains open after birth. This creates a left-to-right shunt of blood from the aorta to the pulmonary artery and results in recirculation of arterial blood through the lungs. Normally, the ductus closes within days to weeks after birth. Failure to close is most prevalent in premature neonates, probably as a result of abnormalities in oxygenation or the relaxant action of prostaglandin E, which prevents ductal spasm and contracture necessary for closure. However, most of the time, the cause of this condition is unknown. PDA commonly accompanies rubella syndrome and may be associated with other congenital defects, such as coarctation of the aorta, VSD, and pulmonary and aortic stenoses.

Initially, PDA may produce no clinical effects, but in time it can precipitate pulmonary vascular disease, causing symptoms to appear by age 40. PDA is found in 1 of every 2,000 infants and is the most common congenital heart defect found in adults. It affects twice as many females as males. Additionally, babies born at above 10,000 feet in altitude are more affected.

The prognosis is good if the shunt is small or surgical repair is effective. Otherwise, PDA may advance to intractable heart failure, which may be fatal. (See *Understanding patent ductus arteriosus*.)

Understanding patent ductus arteriosus

The ductus arteriosus is a fetal blood vessel that connects the pulmonary artery to the descending aorta. Normally, the ductus closes within weeks after birth. However, with patent ductus arteriosus (PDA), it remains open, creating a left-to-right shunt of blood from the aorta to the pulmonary artery and resulting in recirculation of arterial blood through the lungs. PDA is classified as an increased pulmonary blood flow defect.



Pathophysiology

In PDA, relative resistances in pulmonary and systemic vasculature and the size of the ductus determine the amount of left-to-right shunting. The left atrium and left ventricle must accommodate the increased pulmonary venous return, in turn increasing filling pressure and workload on the left side of the heart and possibly causing heart failure. In the final stages of untreated PDA, the left-to-right shunt leads to chronic pulmonary artery hypertension that becomes fixed and unreactive. This causes the shunt to reverse; unoxygenated blood thus enters systemic circulation, causing cyanosis.

Complications

- Left-sided heart failure
- Pulmonary artery hypertension
- Respiratory distress (children)

Signs and symptoms

In neonates, especially those who are premature, a large PDA usually produces respiratory distress, with signs of heart failure due to the tremendous volume of blood shunted to the lungs through a patent ductus and the increased workload on the left side of the heart. Other characteristic features may include heightened susceptibility to respiratory tract infections, slow motor development, and failure to thrive. Most children with PDA have no symptoms except cardiac ones. Others may exhibit signs of heart disease, such as physical underdevelopment, fatigability, and frequent respiratory tract infections. Adults with undetected PDA may develop pulmonary vascular disease and, by age 40, may display fatigability and dyspnea on exertion. About 10% of them also develop infective endocarditis.

Auscultation reveals the classic machinery murmur (Gibson murmur): a continuous murmur (during systole and diastole) best heard at the heart's base, at the second left ICS under the left clavicle in 85% of children with PDA. This murmur may obscure S_2. However, with a right-to-left shunt, such a murmur may be absent. Palpation may reveal a thrill at the left sternal border and a prominent left ventricular impulse. Peripheral arterial pulses are bounding (Corrigan pulse); pulse pressure is widened because of an elevation in systolic blood pressure and, primarily, a drop in diastolic pressure.

Diagnosis

- Chest X-ray may show increased pulmonary vascular markings, prominent pulmonary arteries, and left ventricle and aorta enlargement.
- ECG may be normal or may indicate left atrial or ventricular hypertrophy and, in pulmonary vascular disease, biventricular hypertrophy.
- Echocardiography confirms the diagnosis, detecting and helping to estimate the size of a PDA. It also reveals an enlarged left atrium and left ventricle or right ventricular hypertrophy from pulmonary vascular disease.

CONFIRMING DIAGNOSIS *Cardiac catheterization can also be performed and shows pulmonary arterial oxygen content higher than right ventricular content because of the influx of aortic blood. Increased PAP indicates a large shunt or, if it exceeds systemic arterial pressure, severe pulmonary vascular disease. Catheterization allows calculation of blood volume crossing the ductus and can rule out associated cardiac defects. Dye injection definitively demonstrates PDA.*

Treatment

Asymptomatic infants with PDA require no immediate treatment. Those with heart failure require fluid restriction, diuretics, and cardiac glycosides to minimize or control symptoms. If these measures can't control heart failure, surgery is necessary to ligate the ductus. If symptoms are mild, surgical correction is usually delayed until the infant is between ages 6 months and 3 years, unless problems develop. Before surgery, children with PDA require antibiotics to protect against infective endocarditis.

Other forms of therapy include cardiac catheterization to deposit a plug or coil in the ductus to stop shunting or administration of indomethacin I.V. (a prostaglandin inhibitor that's an alternative to surgery in premature neonates) to induce ductus spasm and closure.

Special considerations

PDA necessitates careful monitoring, patient and family teaching, and emotional support.

- Watch carefully for signs of PDA in all premature neonates.
- Be alert for respiratory distress symptoms resulting from heart failure, which may develop rapidly in a premature neonate. Frequently assess vital signs, ECG, electrolyte levels, and intake and output. Record response to diuretics and other therapy. Watch for signs of digoxin toxicity (poor feeding and vomiting).
- If the infant receives indomethacin for ductus closure, watch for possible adverse effects, such as diarrhea, jaundice, bleeding, and renal dysfunction.
- Before surgery, carefully explain all treatments and tests to parents. Include the child in your explanations. Arrange for the child and her parents to meet the ICU staff. Tell them about expected I.V. lines, monitoring equipment, and postoperative procedures.

- Immediately after surgery, the child may have a CVP catheter and an arterial line in place. Carefully assess vital signs, intake and output, and arterial and venous pressures. Provide pain relief as needed.
- Before discharge, review instructions to the parents about activity restrictions based on the child's tolerance and energy levels. Advise parents not to become overprotective as their child's tolerance for physical activity increases.
- Stress the need for regular follow-up examinations. Advise parents to inform any practitioner who treats their child about the history of surgery for PDA—even if the child is being treated for an unrelated medical problem.

Congenital cyanotic defects

TETRALOGY OF FALLOT

Causes and incidence

Tetralogy of Fallot is a combination of four cardiac defects: VSD, right ventricular outflow tract obstruction (PS), right ventricular hypertrophy, and dextroposition of the aorta, with overriding of the VSD. Blood shunts right to left through the VSD, permitting unoxygenated blood to mix with oxygenated blood, resulting in cyanosis. Tetralogy of Fallot sometimes coexists with other congenital heart defects, such as PDA or ASD.

The cause of tetralogy of Fallot is unknown, but it results from embryologic hypoplasia of the outflow tract of the right ventricle. Multiple factors, such as Down syndrome, have been associated with its presence. Prenatal risk factors include maternal rubella or other viral illnesses, poor prenatal nutrition, maternal alcoholism, mother older than age 40, and diabetes.

Tetralogy of Fallot occurs in about 5 of every 10,000 infants and accounts for about 10% of all congenital heart diseases. It occurs equally in boys and girls. Before surgical advances made correction possible, about one third of these children died in infancy.

Pathophysiology

Tetralogy of Fallot is present at birth.

Pathophysiology depends on the degree of right ventricular outflow obstruction. A mild obstruction may result in a net left-to-right shunt through the VSD; a severe obstruction causes a right-to-left shunt, resulting in low systemic arterial saturation (cyanosis) that is unresponsive to supplemental oxygen.

Each patient may have a varying degree of defect.

Complications

- Cerebral abscess
- Pulmonary thrombosis
- Venous thrombosis
- Cerebral embolism
- Infective endocarditis

Signs and symptoms

Generally, the hallmark of the disorder is cyanosis, which usually becomes evident within several months after birth but may be present at birth if the neonate has severe PS. Between ages 2 months and 2 years, children with tetralogy of Fallot may experience cyanotic or "tet" spells. Such spells result from increased right-to-left shunting, possibly caused by spasm of the right ventricular outflow tract, increased systemic venous return, or decreased systemic arterial resistance.

Exercise, crying, straining, infection, or fever can precipitate blue spells. Blue spells are characterized by dyspnea; deep, sighing respirations; bradycardia; fainting; seizures; and loss of consciousness. Older children may also develop other signs of poor oxygenation, such as clubbing, diminished exercise tolerance, increasing dyspnea on exertion, growth retardation, and eating difficulties. These children habitually squat when they feel short of breath; this is thought to decrease venous return of unoxygenated blood from the legs and increase systemic arterial resistance.

Children with tetralogy of Fallot also risk developing cerebral abscesses, pulmonary thrombosis, venous thrombosis or cerebral embolism, and infective endocarditis.

In females with tetralogy of Fallot who live to childbearing age, the incidence of spontaneous abortion, premature births, and low birth weight rises.

Diagnosis

In a patient with tetralogy of Fallot, auscultation detects a loud systolic heart murmur (best heard along the left sternal border), which may diminish or obscure the pulmonic component of S_2. In a patient with a large PDA, the continuous murmur of the ductus obscures the systolic murmur. Palpation may reveal a cardiac thrill at the left sternal border and an obvious right ventricular impulse. The inferior sternum appears prominent.

The results of special tests also support the diagnosis:

- Chest X-ray may demonstrate decreased pulmonary vascular marking, depending on the pulmonary obstruction's severity, and a boot-shaped cardiac silhouette.
- ECG shows right ventricular hypertrophy, right axis deviation, and, possibly, right atrial hypertrophy.

◆ Echocardiography identifies septal overriding of the aorta, the VSD, and PS and detects the hypertrophied walls of the right ventricle.
◆ Pulse oximetry shows a decrease in oxygen saturation.

CONFIRMING DIAGNOSIS *Cardiac catheterization confirms the diagnosis by visualizing PS, the VSD, and the overriding aorta and ruling out other cyanotic heart defects. This test also measures the degree of oxygen saturation in aortic blood.*

Treatment

Effective management of tetralogy of Fallot necessitates prevention and treatment of complications, measures to relieve cyanosis, and palliative or corrective surgery. During cyanotic spells, the knee–chest position and administration of oxygen and morphine improve oxygenation. Propranolol (a beta-adrenergic blocking agent) may prevent blue spells.

Palliative surgery is performed in infants with potentially fatal hypoxic spells or occasionally needed prior to final correction. The goal of surgery is to enhance blood flow to the lungs to reduce hypoxia; this is often accomplished by joining the subclavian artery to the pulmonary artery (modified Blalock–Taussig procedure). Supportive measures include prophylactic antibiotics to prevent infective endocarditis or cerebral abscess administered before, during, and after bowel, bladder, or any other surgery or dental treatments. Management may also include phlebotomy in children with polycythemia.

Complete corrective surgery to relieve PS and close the VSD, directing left ventricular outflow to the aorta, requires cardiopulmonary bypass with hypothermia to decrease oxygen utilization during surgery, especially in young children. An infant may have this corrective surgery without prior palliative surgery. It's usually done when progressive hypoxia and polycythemia impair the quality of life, rather than at a specific age. However, most children require surgery, some as young as 6 months old as long as oxygen levels remain adequate.

Special considerations

◆ Explain tetralogy of Fallot to the parents. Inform them that their child will set their own exercise limits and will know when to rest. Make sure they understand that their child can engage in physical activity, and advise them not to be overprotective.
◆ Teach the parents to recognize serious hypoxic spells, which can dramatically increase cyanosis; deep, sighing respirations; and loss of consciousness. Tell them to place their child in the knee–chest position and to report such spells immediately. Emergency treatment may be necessary.
◆ Instruct the parents on ways to prevent overexerting their child, such as feeding slowly and providing smaller and more frequent meals. Tell them that remaining calm may decrease anxiety and that anticipating needs may minimize crying. Encourage the parents to recruit other family members in the care of the child to help prevent their own exhaustion.
◆ To prevent infective endocarditis and other infections, warn the parents to keep their child away from people with infections. Urge them to encourage good dental hygiene, and tell them to watch for ear, nose, and throat infections and dental caries, all of which necessitate immediate treatment. When dental care, infections, or surgery requires prophylactic antibiotics, tell the parents to make sure the child completes the prescribed regimen.
◆ If the child requires medical attention for an unrelated problem, advise the parents to inform the practitioner immediately of the child's history of tetralogy of Fallot because any treatment must take this serious heart defect into consideration.
◆ During hospitalization, alert the staff to the child's condition. Because of the right-to-left shunt through the VSD, treat I.V. lines like arterial lines. A clot dislodged from a catheter tip in a vein can cross the VSD and cause cerebral embolism. The same thing can happen if air enters the venous lines.

After palliative surgery:

◆ Monitor oxygenation and arterial blood gas (ABG) values closely in the ICU.
◆ If the child has undergone the modified Blalock–Taussig procedure, don't use the arm on the operative side for measuring blood pressure, inserting I.V. lines, or drawing blood samples, because blood perfusion on this side diminishes greatly until collateral circulation develops. Note this on the child's chart and at the bedside.

After corrective surgery:

◆ Watch for right bundle branch block or more serious disturbances of AV conduction and for ventricular ectopic beats.
◆ Be alert for other postoperative complications, such as bleeding, right-sided heart failure, and respiratory failure. After surgery, transient heart failure is common and may require treatment with digoxin and diuretics.
◆ Monitor left atrial pressure directly. A pulmonary artery catheter may also be used to check central venous and PAPs.
◆ Frequently check color and vital signs. Obtain ABG measurements regularly to assess oxygenation. Suction to prevent atelectasis and

pneumonia, as needed. Monitor mechanical ventilation.

- Monitor and record intake and output accurately.
- If AV block develops with a low heart rate, a temporary external pacemaker may be necessary.
- If blood pressure or cardiac output is inadequate, catecholamines may be ordered by continuous I.V. infusion. To decrease left ventricular workload, administer nitroprusside, if ordered, and provide analgesics, as needed.
- Keep the parents informed about their child's progress. After discharge, the child may require digoxin, diuretics, and other drugs. Stress the importance of complying with the prescribed regimen, and make sure the parents know how and when to administer these medications. Teach the parents to watch for signs of digoxin toxicity (anorexia, nausea, and vomiting). Prophylactic antibiotics to prevent infective endocarditis will still be required. Advise the parents to avoid becoming overprotective as the child's tolerance for physical activity rises.

TRANSPOSITION OF THE GREAT ARTERIES

Causes and incidence

In this congenital heart defect, the great arteries are reversed: the aorta arises from the right ventricle and the pulmonary artery from the left ventricle, producing two noncommunicating circulatory systems (pulmonary and systemic). Transposition accounts for about 3% of all congenital heart defects and often coexists with other congenital heart defects, such as VSD, VSD with PS, ASD, and PDA. It affects two to three times more males than females. Transposition of the great arteries results from faulty embryonic development, but the cause of such development is unknown. Transposition of the great arteries occurs in about 30 of every 100,000 infants.

Pathophysiology

In transposition, oxygenated blood returning to the left side of the heart is carried back to the lungs by a transposed pulmonary artery; unoxygenated blood returning to the right side of the heart is carried to the systemic circulation by a transposed aorta.

Communication between the pulmonary and systemic circulations is necessary for survival. In infants with isolated transposition, blood mixes only at the patent foramen ovale and at the PDA, resulting in slight mixing of unoxygenated systemic blood and oxygenated pulmonary blood. In infants with concurrent cardiac defects, greater mixing of blood occurs.

Complications

- Chronic heart failure
- Poor oxygenation
- Arrhythmias
- Right-sided heart failure

Signs and symptoms

Within the first few hours after birth, neonates with transposition of the great arteries and no other heart defects generally show cyanosis and tachypnea, which worsen with crying. After several days or weeks, such neonates usually develop signs of heart failure (gallop rhythm, tachycardia, dyspnea, hepatomegaly, and cardiomegaly). S_2 is louder than normal because the anteriorly transposed aorta is directly behind the sternum; in many cases, however, no murmur can be heard during the first few days of life. Associated defects (ASD, VSD, or PDA) cause their typical murmurs and may minimize cyanosis but may also cause other complications (especially severe heart failure). VSD with PS produces a characteristic murmur and severe cyanosis.

As infants with this defect grow older, cyanosis is their most prominent abnormality. However, they also develop diminished exercise tolerance, fatigability, coughing, clubbing, and more pronounced murmurs if ASD, VSD, PDA, or PS is present.

Diagnosis

- Chest X-rays are normal in the first days of life. Within days to weeks, right atrial and right ventricular enlargement characteristically cause the heart to appear oblong. X-rays also show increased pulmonary vascular markings, except when PS coexists.
- ECG typically reveals right axis deviation and right ventricular hypertrophy but may be normal in a neonate.

CONFIRMING DIAGNOSIS *Echocardiography demonstrates the reversed position of the aorta and pulmonary artery and records echoes from both semilunar valves simultaneously, due to aortic valve displacement. It also detects other cardiac defects. Cardiac catheterization reveals decreased oxygen saturation in left ventricular blood and aortic blood; increased right atrial, right ventricular, and pulmonary artery oxygen saturation; and right ventricular systolic pressure equal to systemic pressure. Dye injection reveals the transposed vessels and the presence of any other cardiac defects.*

- ABG measurements indicate hypoxia and secondary metabolic acidosis.

Treatment

An infant with transposition may undergo atrial balloon septostomy (Rashkind procedure)

during cardiac catheterization. This procedure enlarges the patent foramen ovale, which improves oxygenation by allowing greater mixing of the pulmonary and systemic circulations. Atrial balloon septostomy requires passage of a balloon-tipped catheter through the foramen ovale and subsequent inflation and withdrawal across the atrial septum. This procedure alleviates hypoxia to a certain degree. Afterward, digoxin and diuretics can lessen heart failure until the infant is ready to withstand corrective surgery (usually by 1 to 2 weeks of age).

One of three surgical procedures can correct transposition, depending on the defect's physiology. The Mustard procedure replaces the atrial septum with a Dacron or pericardial partition that allows systemic venous blood to be channeled to the pulmonary artery—which carries the blood to the lungs for oxygenation—and oxygenated blood returning to the heart to be channeled from the pulmonary veins into the aorta. The Senning procedure accomplishes the same result, using the atrial septum to create partitions to redirect blood flow. In the arterial switch, or Jantene procedure, transposed arteries are surgically anastomosed to the correct ventricle. For this procedure to be successful, the left ventricle must be used to pump at systemic pressure, as it does in neonates or in children with a left ventricular outflow obstruction or a large VSD. The Jantene procedure is the procedure of choice; however, the Mustard and Senning procedures may be used when specific anatomic conditions exist.

Special considerations

- Explain cardiac catheterization and all necessary procedures to the parents. Offer emotional support.
- Monitor vital signs, ABG values, urine output, and CVP, watching for signs of heart failure. Give digoxin and I.V. fluids, being careful to avoid fluid overload.
- Teach the parents to recognize signs of heart failure and digoxin toxicity (poor feeding and vomiting). Stress the importance of regular checkups to monitor cardiovascular status.
- Teach the parents to protect their infant from infection and to give antibiotics.
- Tell the parents to let their child develop normally. They need not restrict activities; let the child set his or her own limits.
- If the patient is scheduled for surgery, explain the procedure to the parents and child, if old enough. Teach them about the ICU and introduce them to the staff. Also explain postoperative care.
- Preoperatively, monitor ABG values, acid–base balance, intake and output, and vital signs.

After corrective surgery:

- Monitor cardiac output by checking blood pressure, skin color, heart rate, urine output, central venous and left atrial pressures, and level of consciousness (LOC). Report abnormalities or changes.
- Carefully monitor ABG levels and report changes in trends.
- To detect supraventricular conduction blocks and arrhythmias, monitor the patient closely. Watch for signs of AV blocks, atrial arrhythmias, and faulty SA function.
- After the Mustard or Senning procedure, watch for signs of baffle obstruction such as marked facial edema.
- Encourage parents to help their child assume new activity levels and independence. Teach them about postoperative antibiotic prophylaxis for endocarditis.

Acquired inflammatory heart disease

MYOCARDITIS

Causes and incidence

Myocarditis is focal or diffuse inflammation of the cardiac muscle (myocardium). It may be acute or chronic and can occur at any age. In many cases, myocarditis fails to produce specific cardiovascular symptoms or electrocardiogram (ECG) abnormalities, and recovery is usually spontaneous, without residual defects. Occasionally, myocarditis is complicated by heart failure; in rare cases, it leads to cardiomyopathy.

Myocarditis may result from:

- bacterial infections—diphtheria; tuberculosis; typhoid fever; tetanus; and staphylococcal, pneumococcal, and gonococcal infections
- chemical poisons—such as chronic alcoholism
- helminthic infections—such as trichinosis
- hypersensitive immune reactions—acute rheumatic fever and postcardiotomy syndrome
- parasitic infections—especially South American trypanosomiasis (Chagas disease) in infants and immunosuppressed adults; also toxoplasmosis
- radiation therapy—large doses of radiation to the chest in treating lung or breast cancer
- viral infections (most common cause in the United States and Western Europe)—coxsackievirus A and B strains and, possibly, poliomyelitis, influenza, rubeola, rubella, and adenoviruses and echoviruses

Myocarditis occurs in 1 to 10 of every 100,000 people in the United States. The median age for this disorder is 42, and incidence is equal between males and females. Children, especially neonates, and persons who are immunocompromised or pregnant (especially pregnant black women) are at higher risk for developing this disorder.

Pathophysiology

The pathophysiology of myocarditis is still being researched, but it is usually caused by a virus, as already mentioned. It results in necrosis of myocardial cells either through direct injury or as a result of an autoimmune reaction of an infectious or toxic process. The extent of the involvement depends on the magnitude of the insult; if it extends to the pericardium, myopericarditis occurs.

Complications

- Arrhythmias
- Thromboembolism
- Chronic valvulitis (when disease results from rheumatic fever)
- Recurrence of disease
- Left-sided heart failure (occasional)
- Cardiomyopathy (rare)

Signs and symptoms

Myocarditis usually causes nonspecific symptoms—such as fatigue, dyspnea, palpitations, and fever—that reflect the accompanying systemic infection. Occasionally, it may produce mild, continuous pressure or soreness in the chest (unlike the recurring, stress-related pain of angina pectoris). Although myocarditis is usually self-limiting, it may induce myofibril degeneration that results in right- and left-sided heart failure, with cardiomegaly, JVD, dyspnea, persistent fever with resting or exertional tachycardia disproportionate to the degree of fever, and supraventricular and ventricular arrhythmias. Sometimes myocarditis recurs or produces chronic valvulitis (when it results from rheumatic fever), cardiomyopathy, arrhythmias, and thromboembolism.

Diagnosis

Patient history commonly reveals recent febrile upper respiratory tract infection, viral pharyngitis, or tonsillitis. Physical examination shows supraventricular and ventricular arrhythmias, S_3 and S_4 gallops, a faint S_1, possibly a murmur of mitral insufficiency (from papillary muscle dysfunction) and, if pericarditis is present, a pericardial friction rub.

Laboratory tests can't unequivocally confirm myocarditis, but the following findings support this diagnosis:

- cardiac enzymes: elevated CK, CK-MB, aspartate aminotransferase, and lactate dehydrogenase levels
- increased white blood cell count and erythrocyte sedimentation rate
- elevated antibody titers (such as antistreptolysin-O titer in rheumatic fever)

CONFIRMING DIAGNOSIS *Endomyocardial biopsy is rarely performed to diagnose myocarditis; the procedure is invasive and costly. A negative biopsy doesn't exclude the diagnosis, and a repeat biopsy may be needed.*

ECG typically shows diffuse ST-segment and T-wave abnormalities as in pericarditis, conduction defects (prolonged PR interval), and other supraventricular arrhythmias. Echocardiography demonstrates some degree of left ventricular dysfunction, and radionuclide scanning may identify inflammatory and necrotic changes characteristic of myocarditis.

Stool and throat cultures may identify bacteria.

Treatment

While myositis is usually self-limiting, treatment may include antibiotics for bacterial infection, modified bed rest to decrease cardiac workload, and careful management of complications. Inotropic support of cardiac function with amrinone, dopamine, or dobutamine may be needed. Heart failure requires restriction of activity to minimize myocardial oxygen consumption, supplemental oxygen therapy, sodium restriction, diuretics to decrease fluid retention, and cardiac glycosides to increase myocardial contractility. However, cardiac glycosides should be administered cautiously because some patients with myocarditis may show a paradoxical sensitivity to even small doses. Arrhythmias necessitate prompt but cautious administration of antiarrhythmics because these drugs depress myocardial contractility. Thromboembolism requires anticoagulation therapy. Treatment with corticosteroids or other immunosuppressants may be used to reduce inflammation, but they haven't been shown to change the progression of myocarditis. Nonsteroidal anti-inflammatory drugs are contraindicated during the acute phase (first 2 weeks) because they increase myocardial damage.

Surgical treatment may include left ventricular assistive devices and extracorporeal membrane oxygenation for support of cardiogenic shock. Cardiac transplantation has been beneficial for giant cell myocarditis.

Special considerations

- Assess cardiovascular status frequently, watching for signs of heart failure, such as dyspnea, hypotension, and tachycardia. Check for changes in cardiac rhythm or conduction.
- Observe for signs of digoxin toxicity (anorexia, nausea, vomiting, blurred vision, and cardiac arrhythmias) and for complicating factors that may potentiate toxicity, such as electrolyte imbalance or hypoxia.
- Stress the importance of bed rest. Assist with bathing, as necessary; provide a bedside commode because this stresses the heart less than using a bedpan. Reassure the patient that activity limitations are temporary. Offer diversional activities that are physically undemanding.
- During recovery, recommend that the patient resume normal activities slowly and avoid competitive sports.

PREVENTION

- *Instruct patient to obtain prompt treatment of causative disorders.*
- *Instruct patient to practice good hygiene, including thorough handwashing.*
- *Tell patient to thoroughly wash and cook food.*

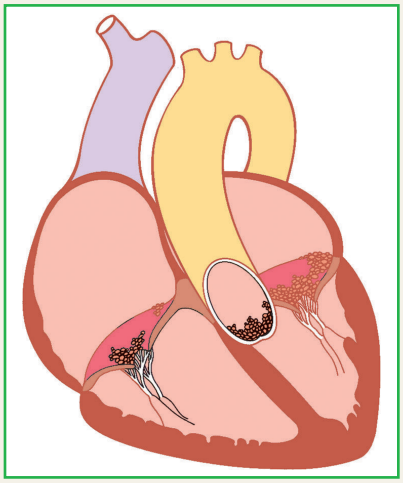
ENDOCARDITIS

Causes and incidence

Endocarditis (also known as *infective* or *bacterial endocarditis*) is an infection of the endocardium, heart valves, or cardiac prostheses resulting from bacterial or fungal invasion. Most cases of endocarditis occur in I.V. drug abusers, patients with prosthetic heart valves, and those with MVP (especially males with a systolic murmur). These conditions have surpassed rheumatic heart disease as the leading risk factor. Other predisposing conditions include coarctation of the aorta, tetralogy of Fallot, subaortic and valvular aortic stenosis, VSDs, PS, Marfan syndrome, degenerative heart disease (especially calcific aortic stenosis), and, rarely, syphilitic aortic valve. However, some patients with endocarditis have no underlying heart disease. In the United States, endocarditis affects 2 to 6 people out of every 100,000. Males are twice as likely as females to acquire this infection, and the mean age of onset is 50. Mortality is associated with increased age, infection of the aortic valve, heart failure and underlying heart disease, and central nervous system complications; mortality rates vary with the infecting organism. Untreated endocarditis is usually fatal, but with proper treatment, 70% of patients recover. The prognosis is worst when endocarditis causes severe valvular damage, leading to insufficiency and heart failure, or when it involves a prosthetic valve.

Degenerative changes in endocarditis

This illustration shows typical vegetations on the endocardium produced by fibrin and platelet deposits on infection sites.



Pathophysiology

The invasion of bacteria or fungi produces vegetative growths on the heart valves, endocardial lining of a heart chamber, or endothelium of a blood vessel that may embolize to the spleen, kidneys, central nervous system, and lungs. In endocarditis, fibrin and platelets aggregate on the valve tissue and engulf circulating bacteria or fungi that flourish and produce friable verrucous vegetations. (See *Degenerative changes in endocarditis*.) Such vegetations may cover the valve surfaces, causing ulceration and necrosis; they may also extend to the chordae tendineae, leading to their rupture and subsequent valvular insufficiency.

Infecting organisms differ depending on the cause of endocarditis. In patients with native valve endocarditis who aren't I.V. drug abusers, causative organisms usually include—in the order of frequency—streptococci (especially *Streptococcus viridans*), staphylococci, or enterococci. Although many other bacteria occasionally cause the disorder, fungal causes are rare in this group. The mitral valve is involved most commonly, followed by the aortic valve.

In patients who are I.V. drug abusers, *Staphylococcus aureus* is the most common infecting organism. Less commonly, streptococci, enterococci, gram-negative bacilli, or fungi cause the

disorder. The tricuspid valve is involved most commonly, followed by the aortic and then the mitral valve.

In patients with prosthetic valve endocarditis, early cases (those that develop within 60 days of valve insertion) are usually due to staphylococcal infection. However, gram-negative aerobic organisms, fungi, streptococci, enterococci, or diphtheroids may also cause the disorder. The course is usually fulminant and is associated with a high mortality. Late cases (occurring after 60 days) present similar to native valve endocarditis.

Complications

- Left-sided heart failure
- Valvular stenosis or insufficiency
- Myocardial erosion

Signs and symptoms

Early clinical features of endocarditis are usually nonspecific and include malaise, weakness, fatigue, weight loss, anorexia, arthralgia, night sweats, chills, valvular insufficiency and, in 90% of patients, an intermittent fever that may recur for weeks. A more acute onset is associated with organisms of high pathogenicity such as *S. aureus.* Endocarditis commonly causes a loud, regurgitant murmur typical of the underlying heart lesion. A suddenly changing murmur or the discovery of a new murmur in the presence of fever is a classic physical sign of endocarditis.

In about 30% of patients, embolization from vegetating lesions or diseased valvular tissue may produce typical features of splenic, renal, cerebral, or pulmonary infarction or of peripheral vascular occlusion:

- splenic infarction—pain in the left upper quadrant, radiating to the left shoulder, and abdominal rigidity
- renal infarction—hematuria, pyuria, flank pain, and decreased urine output
- cerebral infarction—hemiparesis, aphasia, or other neurologic deficits
- pulmonary infarction (most common in right-sided endocarditis, which commonly occurs among I.V. drug abusers and after cardiac surgery)—cough, pleuritic pain, pleural friction rub, dyspnea, and hemoptysis
- peripheral vascular occlusion—numbness and tingling in an arm, leg, finger, or toe, or signs of impending peripheral gangrene

Other signs may include splenomegaly; petechiae of the skin (especially common on the upper anterior trunk) and the buccal, pharyngeal, or conjunctival mucosa; and splinter hemorrhages under the nails. Rarely, endocarditis produces Osler nodes (tender, raised, subcutaneous lesions on the fingers or toes), Roth spots (hemorrhagic areas with white centers on the retina), and Janeway lesions (purplish macules on the palms or soles).

Diagnosis

CONFIRMING DIAGNOSIS *Three or more blood cultures in a 24- to 48-hour period (each from a separate venipuncture) identify the causative organism in up to 90% of patients. Blood cultures should be drawn from three different sites with 1 hour between each draw.*

The remaining 10% may have negative blood cultures, possibly suggesting fungal infection or infections that are difficult to diagnose, such as *Haemophilus parainfluenzae.*

Other abnormal but nonspecific laboratory test results include:

- normal or elevated white blood cell count
- abnormal histiocytes (macrophages)
- elevated erythrocyte sedimentation rate
- normocytic, normochromic anemia (in 70% to 90% of patients)
- proteinuria and microscopic hematuria (in about 50% of patients)
- positive serum rheumatoid factor (in about 50% of patients after endocarditis is present for 3 to 6 weeks)

Echocardiography (particularly, transesophageal) may identify valvular damage; ECG may show atrial fibrillation and other arrhythmias that accompany valvular disease.

Treatment

The goal of treatment is to eradicate the infecting organism with appropriate antimicrobial therapy, which should start promptly and continue over 4 to 6 weeks. Selection of an antibiotic is based on identification of the infecting organism and on sensitivity studies. While awaiting results, or if blood cultures are negative, empiric antimicrobial therapy is based on the likely infecting organism.

Supportive treatment includes bed rest, aspirin for fever and aches, and sufficient fluid intake. Severe valvular damage, especially aortic or mitral insufficiency, may require corrective surgery if refractory heart failure develops, or in cases requiring that an infected prosthetic valve be replaced.

Special considerations

- Before giving antibiotics, obtain a patient history of allergies. Administer antibiotics on time to maintain consistent antibiotic blood levels.
- Observe for signs of infiltration or inflammation at the venipuncture site, possible

PREVENTION

Preventing endocarditis

Any patient who is at risk for or susceptible to endocarditis, such as those with artificial heart valves or other predisposing factors, should have prophylactic antibiotics before dental or other invasive procedures.

In addition, the patient should practice good hygiene, including thoroughly washing hands and washing fruits and vegetables and thoroughly cooking all food to prevent introducing organisms into the system. Maintaining good oral health by daily brushing and flossing and having regular dental checkups can also prevent infection. Be sure to advise the patient to notify the family practitioner as well as the dentist or another specialist that they have a condition that places them at high risk for endocarditis.

complications of long-term I.V. drug administration. To reduce the risk of these complications, rotate venous access sites.

- Watch for signs of embolization (hematuria, pleuritic chest pain, left upper quadrant pain, or paresis), a common occurrence during the first 3 months of treatment. Tell the patient to watch for and report these signs, which may indicate impending peripheral vascular occlusion or splenic, renal, cerebral, or pulmonary infarction.
- Monitor the patient's renal status (blood urea nitrogen [BUN] levels, creatinine clearance, and urine output) to check for signs of renal emboli or evidence of drug toxicity.
- Observe for signs of heart failure, such as dyspnea, tachypnea, tachycardia, crackles, JVD, edema, and weight gain.
- Provide reassurance by teaching the patient and family about this disease and the need for prolonged treatment. Tell them to watch closely for fever, anorexia, and other signs of relapse about 2 weeks after treatment stops. Suggest quiet diversionary activities to prevent excessive physical exertion.
- Make sure susceptible patients understand the need for prophylactic antibiotics before, during, and after dental work, childbirth, and genitourinary, GI, or gynecologic procedures.
- Teach patients how to recognize symptoms of endocarditis and tell them to notify the practitioner at once if such symptoms occur. (See *Preventing endocarditis.*)

PERICARDITIS

Causes and incidence

Pericarditis is an inflammation of the pericardium, the fibroserous sac that envelops, supports, and protects the heart. Common causes of this disease include:

- bacterial, fungal, or viral infection (infectious pericarditis)
- neoplasms (primary or metastatic from lungs, breasts, or other organs)
- high-dose radiation to the chest
- uremia
- hypersensitivity or autoimmune disease, such as acute rheumatic fever (most common cause of pericarditis in children), systemic lupus erythematosus (SLE), and rheumatoid arthritis
- postcardiac injury such as MI, which later causes an autoimmune reaction (Dressler syndrome) in the pericardium; trauma; or surgery that leaves the pericardium intact but causes blood to leak into the pericardial cavity
- drugs, such as hydralazine or procainamide
- idiopathic factors (most common in acute pericarditis)

Less common causes include aortic aneurysm with pericardial leakage and myxedema with cholesterol deposits in the pericardium.

Pericarditis most commonly affects men 20 to 50 years old, but it can also occur in children after infection with an adenovirus or coxsackievirus.

The prognosis depends on the underlying cause but is generally good in acute pericarditis, unless constriction occurs.

Pathophysiology

The pericardium protects the heart mechanically and reduces friction of the surrounding structures through a small amount of pericardial fluid (25 to 50 mL). Inflammation of the layers of the pericardium leads to an increase in the production of this fluid in the form of exudate. Pericarditis occurs in both acute

and chronic forms. Acute pericarditis can be fibrinous or effusive, with purulent serous or hemorrhagic exudate; chronic constrictive pericarditis is characterized by dense fibrous pericardial thickening.

Complications

- Pericardial effusion
- Cardiac tamponade
- Shock
- Cardiovascular collapse
- Death

Signs and symptoms

Acute pericarditis typically produces a sharp and often sudden pain that usually starts over the sternum and radiates to the neck, shoulders, back, and arms. However, unlike the pain of MI, pericardial pain is often pleuritic, increasing with deep inspiration and decreasing when the patient sits up and leans forward, pulling the heart away from the diaphragmatic pleurae of the lungs.

Pericardial effusion, the major complication of acute pericarditis, may produce effects of heart failure (such as dyspnea, orthopnea, and tachycardia), ill-defined substernal chest pain, and a feeling of fullness in the chest. (See *Patterns of cardiac pain.*)

ALERT *If the fluid accumulates rapidly, cardiac tamponade may occur, resulting in pallor, clammy skin, hypotension, pulsus paradoxus (a decrease in systolic blood pressure of 15 mm Hg or more during slow inspiration), JVD and, eventually, cardiovascular collapse and death.*

Chronic constrictive pericarditis causes a gradual increase in systemic venous pressure and produces symptoms similar to those of chronic right-sided heart failure (fluid retention, ascites, and hepatomegaly).

Diagnosis

Because pericarditis commonly coexists with other conditions, the diagnosis of acute pericarditis depends on typical clinical features and elimination of other possible causes.

Patterns of cardiac pain

Although pain perception is individualistic, specific characteristics are associated with different types of cardiac pain, as shown below.

Pericarditis	*Angina*	*Myocardial infarction*
Onset and duration ♦ Sudden onset; continuous pain lasting for days; residual soreness **Location and radiation** ♦ Substernal pain to left of midline; radiation to back or subclavicular area **Quality and intensity** ♦ Mild ache to severe pain, deep or superficial; "stabbing," "knifelike" **Signs and symptoms** ♦ Precordial friction rub; increased pain with movement, inspiration, laughing, coughing; decreased pain with sitting or leaning forward (sitting up pulls heart away from diaphragm) **Precipitating factors** ♦ Myocardial infarction or upper respiratory tract infection; invasive cardiac trauma	**Onset and duration** ♦ Gradual or sudden onset; pain usually lasts <15 minutes and not >30 minutes (average: 3 minutes) **Location and radiation** ♦ Substernal or anterior chest pain, not sharply localized; radiation to back, neck, arms, jaws, even upper abdomen or fingers **Quality and intensity** ♦ Mild-to-moderate pressure; deep sensation; varied pattern of attacks; "tightness," "squeezing," "crushing," "pressure" **Signs and symptoms** ♦ Dyspnea, diaphoresis, nausea, desire to void, belching, apprehension **Precipitating factors** ♦ Exertion, stress, eating, cold or hot and humid weather	**Onset and duration** ♦ Sudden onset; pain lasts 30 minutes to 2 hours; waxes and wanes; residual soreness 1 to 3 days **Location and radiation** ♦ Substernal, midline, or anterior chest pain; radiation to jaws, neck, back, shoulders, or one or both arms **Quality and intensity** ♦ Persistent, severe pressure; deep sensation; "crushing," "squeezing," "heavy," "oppressive" **Signs and symptoms** ♦ Nausea, vomiting, apprehension, dyspnea, diaphoresis, increased or decreased blood pressure; gallop heart sound, "sensation of impending doom" **Precipitating factors** ♦ Occurrence at rest or during physical exertion or emotional stress

The pericardial friction rub, a classic symptom, is a grating sound heard as the heart moves. It can usually be auscultated best during forced expiration, while the patient leans forward or is on hands and knees in bed. It may have up to three components, corresponding to the timing of atrial systole, ventricular systole, and the rapid-filling phase of ventricular diastole. Occasionally, this friction rub is heard only briefly or not at all. Nevertheless, its presence, together with other characteristic features, is diagnostic of acute pericarditis. In addition, if acute pericarditis has caused very large pericardial effusions, physical examination reveals increased cardiac dullness and diminished or absent apical impulse and distant heart sounds.

Chest X-ray, echocardiogram, chest MRI, heart MRI, heart computed tomography scan, and radionuclide scanning can detect fluid that has accumulated in the pericardial sac. They may also show enlargement of the heart and signs of inflammation or scarring, depending on the cause of pericarditis.

In patients with chronic pericarditis, acute inflammation or effusions don't occur—only restricted cardiac filling.

Laboratory results reflect inflammation and may identify its cause:

- normal or elevated white blood cell count, especially in infectious pericarditis
- elevated erythrocyte sedimentation rate
- slightly elevated cardiac enzyme levels with associated myocarditis
- culture of pericardial fluid obtained by open surgical drainage or cardiocentesis (sometimes identifies a causative organism in bacterial or fungal pericarditis)
- ECG showing the following changes in acute pericarditis: elevation of ST segments in the standard limb leads and most precordial leads without the significant changes in QRS morphology that occur with MI, atrial ectopic rhythms such as atrial fibrillation and, in pericardial effusion, diminished QRS voltage

Other pertinent laboratory data include BUN levels to check for uremia, antistreptolysin-O titers to detect rheumatic fever, and a purified protein derivative skin test to check for tuberculosis. In pericardial effusion, echocardiography is diagnostic when it shows an echo-free space between the ventricular wall and the pericardium.

Treatment

The goal of treatment is to relieve symptoms and manage the underlying systemic disease. In acute idiopathic pericarditis and postthoracotomy pericarditis, treatment consists of bed rest as long as fever and pain persist, and nonsteroidal drugs, such as aspirin and indomethacin, to relieve pain and reduce inflammation. Post-MI patients should avoid nonsteroidal anti-inflammatory drugs and steroids because they may interfere with myocardial scar formation. If these drugs fail to relieve symptoms, corticosteroids may be used. Although corticosteroids produce rapid and effective relief, they must be used cautiously because episodes may recur when therapy is discontinued.

Infectious pericarditis that results from disease of the left pleural space, mediastinal abscesses, or septicemia requires antibiotics (possibly by direct pericardial injection), surgical drainage, or both. Cardiac tamponade may require pericardiocentesis. Signs of tamponade include pulsus paradoxus, JVD, dyspnea, and shock.

Recurrent pericarditis may necessitate partial pericardectomy, which creates a "window" that allows fluid to drain into the pleural space. In constrictive pericarditis, total pericardectomy to permit adequate filling and contraction of the heart may be necessary. Treatment must also include management of rheumatic fever, uremia, tuberculosis, and other underlying disorders.

Special considerations

A patient with pericarditis needs complete bed rest. In addition, healthcare includes:

- assessing pain in relation to respiration and body position to distinguish pericardial pain from myocardial ischemic pain
- placing the patient in an upright position to relieve dyspnea and chest pain; providing analgesics and oxygen; and reassuring the patient with acute pericarditis that the condition is temporary and treatable
- monitoring for signs of cardiac compression or cardiac tamponade, possible complications of pericardial effusion (Signs include decreased blood pressure, increased CVP, and pulsus paradoxus. Because cardiac tamponade requires immediate treatment, keep a pericardiocentesis set handy whenever pericardial effusion is suspected.)
- explaining tests and treatments to the patient (If surgery is necessary, the patient should learn deep breathing and coughing exercises beforehand. Postoperative care is similar to that given after cardiothoracic surgery.)

RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE

Causes and incidence

Acute rheumatic fever is a systemic inflammatory disease of childhood, in many cases recurrent, that follows a group A beta-hemolytic

streptococcal infection. Rheumatic heart disease refers to the cardiac manifestations of rheumatic fever and includes pancarditis (myocarditis, pericarditis, and endocarditis) during the early acute phase and chronic valvular disease later. Although rheumatic fever tends to be familial, this may merely reflect contributing environmental factors. For example, in lower socioeconomic groups, incidence is highest in children between 5 and 15 years old, probably as a result of malnutrition and crowded living conditions. This disease strikes generally during cool, damp weather in the winter and early spring. In the United States, it's most common in the northern states. Long-term antibiotic therapy can minimize the recurrence of rheumatic fever, reducing the risk of permanent cardiac damage and eventual valvular deformity. However, severe pancarditis occasionally produces fatal heart failure during the acute phase. Of the patients who survive this complication, about 20% die within 10 years.

Pathophysiology

Rheumatic fever appears to be a hypersensitivity reaction to a group A beta-hemolytic streptococcal infection, in which antibodies manufactured to combat streptococci react and produce characteristic lesions at specific tissue sites, especially in the heart and joints. Because very few persons (0.3%) with streptococcal infections ever contract rheumatic fever, altered host resistance must be involved in its development or recurrence.

Complications

- Destruction of mitral and aortic valves
- Severe pancarditis
- Pericardial effusion
- Fatal heart failure

Signs and symptoms

In 95% of patients, rheumatic fever characteristically follows a streptococcal infection that appeared a few days to 6 weeks earlier. A temperature of at least 100.4° F (38° C) occurs, and most patients complain of migratory joint pain or polyarthritis. Swelling, redness, and signs of effusion usually accompany such pain, which most commonly affects the knees, ankles, elbows, or hips. In 5% of patients (generally those with carditis), rheumatic fever causes skin lesions such as erythema marginatum, a nonpruritic, macular, transient rash that gives rise to red lesions with blanched centers. Rheumatic fever may also produce firm, movable, nontender, subcutaneous nodules about 3 mm to 2 cm in diameter, usually near tendons or bony prominences of joints (especially the elbows, knuckles, wrists, and knees) and less often on the scalp and backs of the hands. These nodules persist for a few days to several weeks and, like erythema marginatum, often accompany carditis.

Later, rheumatic fever may cause transient chorea, which develops up to 6 months after the original streptococcal infection. Mild chorea may produce hyperirritability, a deterioration in handwriting, or an inability to concentrate. Severe chorea (Sydenham chorea) causes purposeless, nonrepetitive, involuntary muscle spasms; poor muscle coordination; and weakness. Chorea always resolves without residual neurologic damage.

The most destructive effect of rheumatic fever is carditis, which develops in up to 50% of patients and may affect the endocardium, myocardium, pericardium, or the heart valves. Pericarditis causes a pericardial friction rub and, occasionally, pain and effusion. Myocarditis produces characteristic lesions called Aschoff bodies (in the acute stages) and cellular swelling and fragmentation of interstitial collagen, leading to the formation of a progressively fibrotic nodule and interstitial scars. Endocarditis causes valve leaflet swelling; erosion along the lines of leaflet closure; and blood, platelet, and fibrin deposits, which form beadlike vegetations. Endocarditis affects the mitral valve most often in females and the aortic valve most often in males. In both females and males, endocarditis affects the tricuspid valves occasionally and the pulmonic valve only rarely.

Severe rheumatic carditis may cause heart failure with dyspnea; right upper quadrant pain; tachycardia; tachypnea; a hacking, nonproductive cough; edema; and significant mitral and aortic murmurs. The most common of such murmurs include:

- a systolic murmur of mitral insufficiency (high-pitched, blowing, holosystolic, loudest at apex, possibly radiating to the anterior axillary line)
- a midsystolic murmur due to stiffening and swelling of the mitral leaflet
- occasionally, a diastolic murmur of aortic insufficiency (low-pitched, rumbling, almost inaudible). Valvular disease may eventually result in chronic valvular stenosis and insufficiency, including mitral stenosis and insufficiency, and aortic insufficiency. In children, mitral insufficiency remains the major sequela of rheumatic heart disease.

Diagnosis

Diagnosis depends on recognition of one or more of the classic symptoms (carditis, rheumatic fever without carditis, polyarthritis, chorea, erythema marginatum, or subcutaneous nodules) and a detailed patient history. Laboratory data support the diagnosis:

- White blood cell count and erythrocyte sedimentation rate may be elevated (during the acute phase); blood studies show slight anemia due to suppressed erythropoiesis during inflammation.
- C-reactive protein is positive (especially during the acute phase).
- Cardiac enzyme levels may be increased in severe carditis.
- Antistreptolysin-O titer is elevated in 95% of patients within 2 months of onset.
- Electrocardiogram changes aren't diagnostic, but PR interval is prolonged in 20% of patients.
- Chest X-rays show normal heart size (except with myocarditis, heart failure, or pericardial effusion).
- Echocardiography helps evaluate valvular damage, chamber size, and ventricular function.
- Cardiac catheterization evaluates valvular damage and left ventricular function in severe cardiac dysfunction.

Treatment

Effective management eradicates the streptococcal infection, relieves symptoms, and prevents recurrence, reducing the chance of permanent cardiac damage. During the acute phase, treatment includes penicillin, sulfadiazine, or erythromycin. Salicylates such as aspirin relieve fever and minimize joint swelling and pain; if carditis is present or salicylates fail to relieve pain and inflammation, corticosteroids may be used. Supportive treatment requires strict bed rest for about 5 weeks during the acute phase with active carditis, followed by a progressive increase in physical activity, depending on clinical and laboratory findings and the response to treatment.

After the acute phase subsides, low-dose antibiotics may be used to prevent recurrence. Such preventive treatment usually continues for 5 years or until age 21 (whichever is longer). Heart failure necessitates continued bed rest and diuretics. Severe mitral or aortic valve dysfunction that causes persistent heart failure requires corrective valvular surgery, including commissurotomy (separation of the adherent, thickened leaflets of the mitral valve), valvuloplasty (inflation of a balloon within a valve), or valve replacement (with prosthetic valve). Such surgery is seldom necessary before late adolescence.

Special considerations

Because rheumatic fever and rheumatic heart disease require prolonged treatment, the care plan should include comprehensive patient teaching to promote compliance with the prescribed therapy.

- Before giving penicillin, ask the patient or parents if the patient has ever had a hypersensitivity reaction to it. If not, warn that such a reaction is possible. Tell them to stop the drug and call the practitioner immediately if the patient develops a rash, fever, chills, or other signs of allergy *at any time* during penicillin therapy.
- Instruct the patient and family to watch for and report early signs of heart failure, such as dyspnea and a hacking, nonproductive cough.
- Stress the need for bed rest during the acute phase, and suggest appropriate, physically undemanding diversions. After the acute phase, encourage family and friends to spend as much time as possible with the patient to minimize boredom. Advise parents to secure tutorial services to help the child keep up with schoolwork during the long convalescence.
- Help the child's parents overcome any guilt feelings they may have about the illness. Tell them that failure to seek treatment for streptococcal infection is common because this illness often seems no worse than a cold. Encourage the child and his or her parents to vent their frustrations during the long, tedious recovery. If the child has severe carditis, help them prepare for permanent changes in lifestyle.
- Teach the patient and his or her family about this disease and its treatment. Warn parents to watch for and immediately report signs of recurrent streptococcal infection—sudden sore throat, diffuse throat redness and oropharyngeal exudate, swollen and tender cervical lymph glands, pain on swallowing, temperature of 101° to 104° F (38.3° to 40° C), headache, and nausea. Urge them to keep the child away from people with respiratory tract infections.
- Promote good dental hygiene to prevent gingival infection. Make sure the patient and his or her family understand the need to comply with prolonged antibiotic therapy and follow-up care and the need for additional antibiotics during dental surgery or procedures. Arrange for a home health nurse to oversee home care if necessary.
- Teach the patient to follow current recommendations of the American Heart Association for prevention of bacterial endocarditis. Antibiotic regimens used to prevent recurrence of acute rheumatic fever are inadequate for preventing bacterial endocarditis.

Valve disorders

VALVULAR HEART DISEASE

Causes and incidence

More than 5 million people in the United States are diagnosed with some form of valvular disease each year. The mitral and aortic valves are most commonly affected. Common causes of each type can be found in *Types of valvular heart disease.*

Pathophysiology

In valvular heart disease, three types of mechanical disruption can occur: stenosis, or narrowing, of the valve opening; incomplete closure of the valve; and prolapse of the valve. A combination of these three in the same valve may also occur. They can result from such disorders as endocarditis (most common), congenital defects, and inflammation, and they can lead to heart failure.

Valvular heart disease occurs in varying forms, described in the following.

- Mitral insufficiency: In this form, blood from the left ventricle flows back into the left atrium during systole, causing the atrium to enlarge to accommodate the backflow. As a result, the left ventricle also dilates to accommodate the increased volume of blood from the atrium and to compensate for diminishing cardiac output. Ventricular hypertrophy and increased end-diastolic pressure result in increased PAP, eventually leading to left- and right-sided heart failure.
- Mitral stenosis: Narrowing of the valve by valvular abnormalities, fibrosis, or calcification obstructs blood flow from the left atrium to the left ventricle. Consequently, left atrial volume and pressure rise and the chamber dilates. Greater resistance to blood flow causes pulmonary hypertension, right ventricular hypertrophy, and right-sided heart failure. Also, inadequate filling of the left ventricle produces low cardiac output.
- Mitral valve prolapse: One or both valve leaflets protrude into the left atrium. *MVP* is the term used when the anatomic prolapse is accompanied by signs and symptoms unrelated to the valvular abnormality.
- Aortic insufficiency: Blood flows back into the left ventricle during diastole, causing fluid overload in the ventricle, which dilates and hypertrophies. The excess volume causes fluid overload in the left atrium, and, finally, the pulmonary system. Left-sided heart failure and pulmonary edema eventually result.
- Aortic stenosis: Increased left ventricular pressure tries to overcome the resistance of the narrowed valvular opening. The added workload increases the demand for oxygen, whereas diminished cardiac output causes poor coronary artery perfusion, ischemia of the left ventricle, and left-sided heart failure.
- Pulmonic insufficiency: Blood ejected into the pulmonary artery during systole flows back into the right ventricle during diastole, causing fluid overload in the ventricle, ventricular hypertrophy and, finally, right-sided heart failure.
- Pulmonic stenosis: Obstructed right ventricular outflow causes right ventricular hypertrophy, eventually resulting in right-sided heart failure.
- Tricuspid insufficiency: Blood flows back into the right atrium during systole, decreasing blood flow to the lungs and the left side of the heart. Cardiac output also lessens. Fluid overload in the right side of the heart can eventually lead to right-sided heart failure.
- Tricuspid stenosis: Obstructed blood flow from the right atrium to the right ventricle causes the right atrium to dilate and hypertrophy. Eventually, this leads to right-sided heart failure and increases pressure in the vena cava.

Treatment

Treatment depends on the nature and severity of associated symptoms. For example, heart failure requires diuretics, a sodium-restricted diet, and, in acute cases, oxygen. Other measures may include anticoagulant therapy or antiplatelet medications to prevent thrombus formation around diseased or replaced valves, prophylactic antibiotics before and after surgery, and valvuloplasty. An IABP may be used temporarily to reduce backflow by enhancing forward blood flow into the aorta.

If the patient has severe signs and symptoms that can't be managed medically, open heart surgery using cardiopulmonary bypass for valve repair or replacement is indicated. Newer procedures are available, such as transcatheter aortic valve replacement, and may be an option, as well. This is a minimally invasive procedure that wedges a replacement valve in the position of the old valve. This valve begins to take over the duties of the old valve while pushing the leaflets of the old valve away.

Special considerations

- Watch closely for signs of heart failure or pulmonary edema and for adverse effects of drug therapy.
- Teach the patient about diet restrictions, medications, and the importance of consistent follow-up care.
- If the patient undergoes surgery, watch for hypotension, arrhythmias, and thrombus

Types of valvular heart disease

Causes and incidence	*Signs and symptoms*	*Diagnostic measures*
Aortic insufficiency		
♦ Results from rheumatic fever, syphilis, hypertension, endocarditis, or may be idiopathic ♦ Associated with Marfan syndrome ♦ Most common in males ♦ Associated with ventricular septal defect, even after surgical closure	♦ Dyspnea, cough, fatigue, palpitations, angina, syncope ♦ Pulmonary venous congestion, heart failure, pulmonary edema (left-sided heart failure), "pulsating" nail beds ♦ Rapidly rising and collapsing pulses (pulsus bisferiens), cardiac arrhythmias, wide pulse pressure in severe insufficiency ♦ Auscultation: reveals S_3 and diastolic blowing murmur at left sternal border ♦ Palpation and visualization of apical impulse in chronic disease	♦ Cardiac catheterization: reduction in arterial diastolic pressures, aortic insufficiency, other valvular abnormalities, and increased left ventricular end-diastolic pressure ♦ X-ray: left ventricular enlargement, pulmonary vein congestion ♦ Echocardiography: left ventricular enlargement, alterations in mitral valve movement (indirect indication of aortic valve disease), and mitral thickening ♦ Electrocardiography (ECG): sinus tachycardia, left ventricular hypertrophy, and left atrial hypertrophy in severe disease
Aortic stenosis		
♦ Results from congenital aortic bicuspid valve (associated with coarctation of the aorta), congenital stenosis of valve cusps, rheumatic fever, or atherosclerosis in elderly persons ♦ Most common in males	♦ Dyspnea on exertion, paroxysmal nocturnal dyspnea, fatigue, syncope, angina, palpitations ♦ Pulmonary venous congestion, heart failure, pulmonary edema ♦ Diminished carotid pulses, decreased cardiac output, cardiac arrhythmias; may have pulsus alternans ♦ Auscultation: reveals systolic murmur at base or in carotids and, possibly, S_4	♦ Cardiac catheterization: pressure gradient across valve (indicating obstruction), increased left ventricular end-diastolic pressures ♦ X-ray: valvular calcification, left ventricular enlargement, and pulmonary venous congestion ♦ Echocardiography: thickened aortic valve and left ventricular wall ♦ ECG: left ventricular hypertrophy
Mitral insufficiency		
♦ Results from rheumatic fever, hypertrophic cardiomyopathy, mitral valve prolapse, myocardial infarction, severe left-sided heart failure, or ruptured chordae tendineae ♦ Associated with other congenital anomalies such as transposition of the great arteries ♦ Rare in children without other congenital anomalies	♦ Orthopnea, dyspnea, fatigue, angina, palpitations ♦ Peripheral edema, jugular vein distention (JVD), hepatomegaly (right-sided heart failure) ♦ Tachycardia, crackles, pulmonary edema ♦ Auscultation: reveals holosystolic murmur at apex, possible split S_2, and S_3	♦ Cardiac catheterization: mitral insufficiency with increased left ventricular end-diastolic volume and pressure, increased atrial pressure and pulmonary artery wedge pressure (PAWP); and decreased cardiac output ♦ X-ray: left atrial and ventricular enlargement, pulmonary venous congestion ♦ Echocardiography: abnormal valve leaflet motion, left atrial enlargement ♦ ECG: left atrial and ventricular hypertrophy, sinus tachycardia, and atrial fibrillation

Types of valvular heart disease (*continued*)

Causes and incidence	*Signs and symptoms*	*Diagnostic measures*
Mitral stenosis		
♦ Results from rheumatic fever (most common cause) ♦ Most common in females ♦ May be associated with other congenital anomalies	♦ Dyspnea on exertion, paroxysmal nocturnal dyspnea, orthopnea, weakness, fatigue, palpitations ♦ Peripheral edema, JVD, ascites, hepatomegaly (right-sided heart failure in severe pulmonary hypertension) ♦ Crackles, cardiac arrhythmias (atrial fibrillation), signs of systemic emboli ♦ Auscultation: reveals loud S_1 or opening snap and diastolic murmur at apex	♦ Cardiac catheterization: diastolic pressure gradient across valve; elevated left atrial pressure and PAWP (>15 mm Hg) with severe pulmonary hypertension and pulmonary artery pressures (PAPs); elevated right-sided heart pressure; decreased cardiac output; and abnormal contraction of the left ventricle ♦ X-ray: left atrial and ventricular enlargement, enlarged pulmonary arteries, and mitral valve calcification ♦ Echocardiography: thickened mitral valve leaflets, left atrial enlargement ♦ ECG: left atrial hypertrophy, atrial fibrillation, right ventricular hypertrophy, and right axis deviation
Mitral valve prolapse syndrome		
♦ Can be genetic or associated with conditions such as Ehlers–Danlos syndrome, Marfan syndrome, Graves disease, and muscular dystrophy ♦ Most commonly affects young women but may occur in both sexes and in all age groups	♦ May produce no signs ♦ Chest pain, palpitations, headache, fatigue, exercise intolerance, dyspnea, light-headedness, syncope, mood swings, anxiety, panic attacks ♦ Auscultation: typically reveals mobile, midsystolic click, with or without mid-to-late systolic murmur	♦ Two-dimensional echocardiography: prolapse of mitral valve leaflets into left atrium ♦ Color-flow Doppler studies: mitral insufficiency ♦ Resting ECG: ST-segment changes, biphasic or inverted T waves in leads II, III, or AV ♦ Exercise ECG: evaluates chest pain and arrhythmias
Pulmonic insufficiency		
♦ May be congenital or may result from pulmonary hypertension ♦ May rarely result from prolonged use of pressure-monitoring catheter in the pulmonary artery	♦ Dyspnea, weakness, fatigue, chest pain ♦ Peripheral edema, JVD, hepatomegaly (right-sided heart failure) ♦ Auscultation: reveals diastolic murmur in pulmonic area	♦ Cardiac catheterization: pulmonic insufficiency, increased right ventricular pressure, and associated cardiac defects ♦ X-ray: right ventricular and pulmonary arterial enlargement ♦ ECG: right ventricular or right atrial enlargement
Pulmonic stenosis		
♦ Results from congenital stenosis of valve cusp or rheumatic heart disease (infrequent) ♦ Associated with other congenital heart defects such as tetralogy of Fallot	♦ Asymptomatic or symptomatic with dyspnea on exertion, fatigue, chest pain, syncope ♦ May lead to peripheral edema, JVD, hepatomegaly (right-sided heart failure) ♦ Auscultation: reveals systolic murmur at left sternal border, split S_2 with delayed or absent pulmonic component	♦ Cardiac catheterization: increased right ventricular pressure, decreased PAP, and abnormal valve orifice ♦ ECG: may show right ventricular hypertrophy, right axis deviation, right atrial hypertrophy, and atrial fibrillation

(*continued*)

Types of valvular heart disease (*continued*)

Causes and incidence	*Signs and symptoms*	*Diagnostic measures*
Tricuspid insufficiency		
♦ Results from right-sided heart failure, rheumatic fever and, rarely, trauma and endocarditis ♦ Associated with congenital disorders ♦ Associated with I.V. drug abuse and infective endocarditis manifesting as tricuspid valve disease	♦ Dyspnea and fatigue ♦ May lead to peripheral edema, JVD, hepatomegaly, and ascites (right-sided heart failure) ♦ Auscultation: reveals possible S_3 and systolic murmur at lower left sternal border that increases with inspiration	♦ Right-sided heart catheterization: high atrial pressure, tricuspid insufficiency, decreased or normal cardiac output ♦ X-ray: right atrial dilation, right ventricular enlargement ♦ Echocardiography: shows systolic prolapse of tricuspid valve, right atrial enlargement ♦ ECG: right atrial or right ventricular hypertrophy, atrial fibrillation
Tricuspid stenosis		
♦ Results from rheumatic fever ♦ May be congenital ♦ Associated with mitral or aortic valve disease ♦ Most common in women	♦ May be symptomatic with dyspnea, fatigue, syncope ♦ Possibly peripheral edema, JVD, hepatomegaly, and ascites (right-sided heart failure) ♦ Auscultation: reveals diastolic murmur at lower left sternal border that increases with inspiration	♦ Cardiac catheterization: increased pressure gradient across valve, increased right atrial pressure, decreased cardiac output ♦ X-ray: right atrial enlargement ♦ Echocardiography: leaflet abnormality, right atrial enlargement ♦ ECG: right atrial hypertrophy, right or left ventricular hypertrophy, and atrial fibrillation

formation. Monitor vital signs, ABG values, intake, output, daily weight, blood chemistries, chest X-rays, and pulmonary artery catheter readings.

Degenerative cardiovascular disorders

HYPERTENSION

Causes and incidence

Hypertension, an intermittent or sustained elevation in diastolic or systolic blood pressure, occurs as two major types: essential (idiopathic) hypertension, the most common, and secondary hypertension, which results from renal disease or another identifiable cause. Malignant hypertension is a severe, fulminant form of hypertension common to both types. Hypertension is a major cause of stroke, cardiac disease, and renal failure. Hypertension affects 25% of adults in the United States. If untreated, it carries a high mortality. Risk factors for hypertension include family history, race (most common in blacks), stress, obesity, a diet high in saturated fats or sodium, tobacco use, sedentary lifestyle, and aging.

Secondary hypertension may result from renal vascular disease; pheochromocytoma; primary hyperaldosteronism; Cushing syndrome; thyroid, pituitary, or parathyroid dysfunction; coarctation of the aorta; pregnancy; neurologic disorders; and use of hormonal contraceptives or other drugs, such as cocaine, epoetin alfa (erythropoietin), and cyclosporine.

The prognosis is good if this disorder is detected early and treatment begins before complications develop. Severely elevated blood pressure (hypertensive crisis) may be fatal. (See *What happens in a hypertensive crisis*, page 31.)

Pathophysiology

Cardiac output and PVR determine blood pressure. Increased blood volume, cardiac rate, and stroke volume as well as arteriolar vasoconstriction can raise blood pressure. The link to sustained hypertension, however, is unclear.

PATHOPHYSIOLOGY

What happens in a hypertensive crisis

Hypertensive crisis is a severe rise in arterial blood pressure caused by a disturbance in one or more of the regulating mechanisms. If left untreated, hypertensive crisis may result in renal, cardiac, or cerebral complications and, possibly, death.

Causes of hypertensive crisis

- Abnormal renal function
- Hypertensive encephalopathy
- Intracerebral hemorrhage
- Heart failure
- Withdrawal of anti-hypertensive drugs (abrupt)
- Myocardial ischemia
- Eclampsia
- Pheochromocytoma
- Monoamine oxidase inhibitor interactions

↓

Prolonged hypertension

↓

Inflammation and necrosis of arterioles

↓

Narrowing of blood vessels

↓

Restriction of blood flow to major organs

↓

Organ damage

↓

Renal

- Decreased renal perfusion
- Progressive deterioration of nephrons
- Decreased ability to concentrate urine
- Increased serum creatinine and blood urea nitrogen levels
- Increased renal tubule permeability with protein leakage into tubules
- Renal insufficiency
- Uremia
- Renal failure

Cardiac

- Decreased cardiac perfusion
- Coronary artery disease
- Angina or myocardial infarction
- Increased cardiac workload
- Left ventricular hypertrophy
- Heart failure

Cerebral

- Decreased cerebral perfusion
- Increased stress on vessel wall
- Arterial spasm
- Ischemia
- Transient ischemic attacks
- Weakening of vessel intima
- Aneurysm formation
- Intracranial hemorrhage

Hypertension may also result from failure of intrinsic regulatory mechanisms:

- Renal hypoperfusion causes release of renin, which is converted by angiotensinogen, a liver enzyme, to angiotensin I. Angiotensin I is converted to angiotensin II, a powerful vasoconstrictor. The resulting vasoconstriction increases afterload. Angiotensin II stimulates adrenal secretion of aldosterone, which increases sodium reabsorption. Hypertonic-stimulated release of antidiuretic hormone from the pituitary gland follows, increasing water reabsorption, plasma volume, cardiac output, and blood pressure.
- Autoregulation changes an artery's diameter to maintain perfusion despite fluctuations in systemic blood pressure. The intrinsic mechanisms responsible include stress relaxation (vessels gradually dilate when blood pressure rises to reduce peripheral resistance) and capillary fluid shift (plasma moves between vessels and extravascular spaces to maintain intravascular volume).
- When the blood pressure drops, baroreceptors in the aortic arch and carotid sinuses decrease their inhibition of the medulla's vasomotor center, which increases sympathetic stimulation of the heart by norepinephrine. This, in turn, increases cardiac output by strengthening the contractile force, increasing the heart rate, and augmenting peripheral resistance by vasoconstriction. Stress can also stimulate the sympathetic nervous system to increase cardiac output and PVR.

Complications

- Stroke
- Coronary artery disease
- Angina
- Myocardial infarction
- Heart failure
- Arrhythmias
- Sudden death
- Cerebral infarction
- Hypertensive encephalopathy
- Hypertensive retinopathy
- Renal failure

Signs and symptoms

Hypertension usually doesn't produce clinical effects until vascular changes in the heart, brain, or kidneys occur. Severely elevated blood pressure damages the intima of small vessels, resulting in fibrin accumulation in the vessels, development of local edema and, possibly, intravascular clotting. Symptoms produced by this process depend on the location of the damaged vessels:

- brain—stroke
- retina—blindness
- heart—myocardial infarction
- kidneys—proteinuria, edema, and, eventually, renal failure

Hypertension increases the heart's workload, causing left ventricular hypertrophy and, later, left- and right-sided heart failure and pulmonary edema.

Classifying blood pressure readings

The Eighth Joint National Committee (JNC8) released updated guidelines in 2014 for classifying and treating hypertension.

The following categories are based on the average of two or more readings taken on separate visits after an initial screening. They apply to adults 18 years old and older.

Normal blood pressure with respect to cardiovascular risk is a systolic reading below 120 mm Hg and a diastolic reading below 80 mm Hg. Historically, hypertension was defined as a systolic blood pressure of 140 mm Hg or higher or a diastolic pressure above 90 mm Hg. The latest guidelines, however, classify hypertension as a systolic reading of 130 mm Hg or higher, or a diastolic pressure above 90 mm Hg.

In addition to classifying stages of hypertension based on average blood pressure readings, clinicians should also take note of target organ disease and any additional risk factors.

Category	*Systolic (mm Hg)*		*Diastolic (mm Hg)*
Normal	<120	and	<80
Elevated	120 to 129	and	<80
Hypertension			
Stage 1	130 to 139	or	80 to 89
Stage 2	≥140	or	≥90
Hypertensive crisis	≥180	and/or	≥120

Diagnosis

Serial blood pressure measurements are obtained and compared to previous readings and trends to reveal an increase in diastolic and systolic pressures. (See *Classifying blood pressure readings*, page 32.)

Auscultation may reveal bruits over the abdominal aorta and the carotid, renal, and femoral arteries; ophthalmoscopy reveals arteriovenous nicking and, in hypertensive encephalopathy, papilledema. Patient history and the following additional tests may show predisposing factors and help identify an underlying cause such as renal disease:

- Urinalysis: Protein levels and red and white blood cell counts may indicate glomerulonephritis.
- Excretory urography: Renal atrophy indicates chronic renal disease; one kidney more than $^{5}/_{8}$″ (1.5 cm) shorter than the other suggests unilateral renal disease.
- Serum potassium: Levels less than 3.5 mEq/L may indicate adrenal dysfunction (primary hyperaldosteronism).
- BUN and serum creatinine: BUN level that's normal or elevated to more than 20 mg/dL and serum creatinine level that's normal or elevated to more than 1.5 mg/dL suggest renal disease.

Other tests help detect cardiovascular damage and other complications:

- ECG may show left ventricular hypertrophy or ischemia.
- Chest X-ray may show cardiomegaly.
- Echocardiography may show left ventricular hypertrophy.

Treatment

The JNC8 recommends the following approach for treating primary hypertension:

- First, help the patient start needed lifestyle modifications, including weight reduction, moderation of alcohol intake, regular physical exercise, reduction in sodium intake, and smoking cessation.
- If the patient fails to achieve the desired blood pressure or make significant progress, continue lifestyle modifications and begin drug therapy.
- Pharmacologic therapy should begin when blood pressure is 140/90 in patients less than 60, and 150/90 in those 60 and older.
- If the patient has comorbid conditions such as diabetes mellitus or chronic kidney disease (CKD), the goal should be to achieve blood pressure less than 140/90 regardless of age.
- In nonblack patients without CKD, consider using a thiazide diuretic, an angiotensin-converting enzyme (ACE) inhibitor, an angiotensin receptor blocker (ARB), or a calcium channel blocker (CCB), alone or in combination.
- In black patients without CKD, consider using a thiazide diuretic or a CCB, alone or in combination.
- In all patients with CKD, consider initiating an ACE or ARB, alone or in combination with another class.
- If the patient has one or more compelling indications, base drug treatment on benefits from outcome studies or existing clinical guidelines. Treatment may include the following, depending on indication:
  - Heart failure—ACE/ARB + beta-adrenergic blocker (BB) + diuretic + spironolactone
  - CAD—ACE, BB, diuretic, CCB
  - Diabetes—ACE/ARB, CCB, diuretic
  - CKD—ACE inhibitor or ARB
  - Postmyocardial infarction/clinical CAD—ACE/ARB + BB
  - Recurrent stroke prevention—ACE, diuretic
  - Pregnancy—labetalol (first line), nifedipine, methyldopa

  Give other antihypertensive drugs as needed.
- If the patient fails to achieve the desired blood pressure, continue lifestyle modifications and optimize drug dosages or add drugs until the goal blood pressure is achieved. Also, consider consultation with a hypertension specialist.

Treatment of secondary hypertension focuses on correcting the underlying cause and controlling hypertensive effects.

Typically, hypertensive emergencies require parenteral administration of a vasodilator or an adrenergic inhibitor. Oral administration of a selected drug, such as nicardipine, hydralazine, or esmolol to rapidly reduces blood pressure. The initial goal is to reduce mean arterial blood pressure by no more than 25% (within minutes to hours) and then to 160/110 mm Hg within 2 hours while avoiding excessive falls in blood pressure that can precipitate renal, cerebral, or myocardial ischemia.

Examples of hypertensive emergencies include hypertensive encephalopathy, intracranial hemorrhage, acute left-sided heart failure with pulmonary edema, and dissecting aortic aneurysm. Hypertensive emergencies are also associated with eclampsia or severe gestational hypertension, unstable angina, and acute MI.

Hypertension without accompanying symptoms or target organ disease seldom requires emergency drug therapy.

Special considerations

◆ To encourage adherence to antihypertensive therapy, suggest that the patient establish a daily routine for taking medication. Warn that uncontrolled hypertension may cause stroke and heart attack. Tell the patient to report adverse drug effects. Also, advise the patient to avoid high-sodium antacids and over-the-counter cold and sinus medications, which contain harmful vasoconstrictors.

◆ Encourage a change in dietary habits. Help the obese patient plan a weight-reduction diet; tell the patient to avoid high-sodium foods (pickles, potato chips, canned soups, and cold cuts) and table salt.

◆ Help the patient examine and modify lifestyle (e.g., by reducing stress and exercising regularly).

◆ If a patient is hospitalized with hypertension, find out if the patient was taking his or her prescribed medication. If not, ask why. If the patient can't afford the medication, refer to appropriate social service agencies. Tell the patient and family to keep a record of drugs used in the past, noting especially those that were or weren't effective. Suggest that the patient record this information on a card and show it to his or her practitioner.

◆ When routine blood pressure screening reveals elevated pressure, first make sure the cuff size is appropriate for the patient's upper arm circumference. Take the pressure in both arms in lying, sitting, and standing positions. Ask the patient if he or she smoked, drank a beverage containing caffeine, or was emotionally upset before the test. Advise the patient to return for

PREVENTION

Preventing hypertension

Certain risk factors for hypertension can't be changed, such as family history, race, and aging, but lifestyle modifications can help prevent hypertension. Based on American Heart Association recommendations, advise your patient to do the following:

Maintain a healthy weight

Maintain a normal weight or lose weight if overweight. Weight loss lowers blood pressure.

Reduce salt

Salt intake should be reduced to about 1.5 g/day. Reducing salt intake can lower blood pressure in individuals with and without hypertension.

Increase potassium

Patients should eat 8 to 10 servings of fruits and vegetables per day to increase potassium intake. Potassium reduces blood pressure in individuals with and without hypertension. Those with kidney disease or heart failure should contact their practitioner before increasing their potassium intake.

Limit alcohol intake

Studies have shown a correlation between alcohol intake and increased blood pressure, especially in individuals who drink >2 drinks/day.

Include exercise

Regular physical activity is defined by the American Heart Association as moderate-intensity exercise such as brisk walking for 150 minutes each week. A lack of physical activity can lead to obesity and increase the risk of hypertension, heart attack, and stroke.

Manage stress

Stress can lead to increased alcohol consumption, smoking, overeating, and other activities that increase the risk of heart attack or stroke. Daily relaxation for short periods during the workday and on weekends can also lower blood pressure.

Stop smoking

Smoking even filtered and light or ultra cigarettes can lead to atherosclerosis. Quitting or not starting is the only way to prevent this major risk factor for heart attack and stroke.

Follow the DASH diet

The Dietary Approaches to Stop Hypertension (DASH) diet encourages vegetables, fruits, and low-fat dairy as well as whole grains, fish, poultry, and nuts. Discourage the eating of fats, red meat, sweets, and sugar-containing beverages. However, individuals with reduced kidney function should always consult their practitioners before starting this diet; it's rich in potassium, which isn't recommended for individuals with these disorders.

blood pressure testing at frequent and regular intervals.

◆ To help identify hypertension and prevent untreated hypertension, participate in public education programs dealing with hypertension and ways to reduce risk factors. Encourage public participation in blood pressure screening programs. Routinely screen all patients, especially those at risk (blacks and people with family histories of hypertension, stroke, or heart attack). (See *Preventing hypertension*, page 34.)

CORONARY ARTERY DISEASE

Causes and incidence

CAD occurs when the arteries that supply blood to the heart muscle harden and narrow, usually as a result of atherosclerosis. The result is the loss of oxygen and nutrients to myocardial tissue because of diminished coronary blood flow. This reduction in blood flow can also lead to coronary syndrome (angina or MI). (See *Understanding coronary artery disease*.)

CAD has been linked to many risk factors: family history, male gender, age (risk increased in those 65 years old or older), hypertension, obesity, smoking, diabetes mellitus, stress, sedentary lifestyle, high serum cholesterol (particularly high low-density lipoprotein cholesterol) or triglyceride levels, low high-density lipoprotein cholesterol levels, high blood homocysteine levels, menopause and, possibly, infections producing inflammatory responses in the artery walls.

Uncommon causes of reduced coronary artery blood flow include dissecting aneurysms, infectious vasculitis, syphilis, and congenital defects in the coronary vascular system. Coronary artery spasms may also impede blood flow. (See *Coronary artery spasm*, page 36.)

CAD is the leading cause of death in the United States. According to the American Heart Association, 1 in 3 deaths is due to cardiovascular disease, and someone dies from such an event about every 40 seconds.

Pathophysiology

In atherosclerosis, a form of arteriosclerosis, fatty, fibrous plaques, possibly including calcium deposits, narrow the lumen of the coronary arteries and reduce the volume of blood that can flow through them, and leading to myocardial ischemia. Plaque formation also predisposes to thrombosis, which can provoke MI.

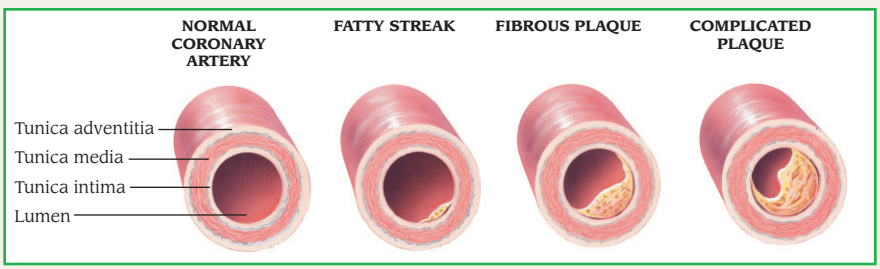
Atherosclerosis usually develops in high-flow, high-pressure arteries, such as those in the heart, brain, kidneys, and in the aorta, especially at bifurcation points.

Signs and symptoms

The classic symptom of CAD is angina, the direct result of inadequate oxygen flow to the myocardium. Anginal pain is usually described as a burning, squeezing, or tight feeling in the substernal or precordial chest that may radiate to the left arm, neck, jaw, or shoulder blade. Typically, the patient clenches a fist over his chest or rubs the left arm when describing the pain, which may be accompanied by nausea, vomiting, fainting, sweating, and cool extremities. Anginal episodes most often follow physical exertion but may also follow emotional excitement, exposure to cold, or a large meal. Some patients, particularly those with diabetes, may not experience typical anginal pain but

Understanding coronary artery disease

Coronary artery disease (CAD) results as atherosclerotic plaque fills the lumens of the coronary arteries and obstructs blood flow. The primary effect of CAD is a diminished supply of oxygen and nutrients to myocardial tissues.



Coronary artery spasm

In coronary artery spasm, a spontaneous, sustained contraction of one or more coronary arteries causes ischemia and dysfunction of the heart muscle. This disorder also causes Prinzmetal angina and even myocardial infarction in patients with unoccluded coronary arteries. Its cause is unknown but possible contributing factors include:

- altered flow of calcium into the cell
- intimal hemorrhage into the medial layer of the blood vessel
- hyperventilation
- elevated catecholamine levels
- fatty buildup in lumen

Signs and symptoms

The major symptom of coronary artery spasm is angina. However, unlike classic angina, this pain often occurs spontaneously and may not be related to physical exertion or emotional stress; it's also more severe, usually lasts longer, and may be cyclic, frequently recurring every day at the same time. Such ischemic episodes may cause arrhythmias, altered heart rate, lower blood pressure and, occasionally, fainting due to diminished cardiac output. Spasm in the left coronary artery may result in mitral insufficiency, producing a loud systolic murmur and, possibly, pulmonary edema, with dyspnea, crackles, hemoptysis, or sudden death.

Treatment

After diagnosis by coronary angiography and electrocardiography (ECG), the patient may receive calcium channel blockers (CCBs; verapamil, nifedipine, or diltiazem) to reduce coronary artery spasm and vascular resistance and nitrates (nitroglycerin or isosorbide dinitrate) to relieve chest pain.

When caring for a patient with coronary artery spasm, explain all necessary procedures and teach them how to take medications safely. For CCB therapy, monitor blood pressure, pulse rate, and ECG patterns to detect arrhythmias. In patients receiving nifedipine and verapamil along with digoxin, monitor digoxin levels and check for signs of digoxin toxicity. Because nifedipine may cause peripheral and periorbital edema, watch for fluid retention.

Because coronary artery spasm is commonly associated with atherosclerotic disease, advise the patient to stop smoking, avoid overeating, maintain a low-fat diet, use alcohol sparingly, and maintain a balance between exercise and rest.

may have dyspnea, fatigue, diaphoresis, or more vague symptoms.

Angina has four major forms: *stable* (pain is predictable in frequency and duration and can be relieved with nitrates and rest), *unstable* (pain increases in frequency and duration and is more easily induced), *Prinzmetal* or *variant* (from unpredictable coronary artery spasm), and *microvascular* (in which impairment of vasodilator reserve causes angina-like chest pain in a patient with normal coronary arteries). Severe and prolonged anginal pain generally suggests MI, with potentially fatal arrhythmias and mechanical failure.

Diagnosis

The patient history—including the frequency and duration of angina and the presence of associated risk factors—is crucial in evaluating CAD. Additional diagnostic measures include the following:

- Electrocardiogram (ECG) during angina may show ischemia and, possibly, arrhythmias such as premature ventricular contractions. ECG is apt to be normal when the patient is pain-free. Arrhythmias may occur without infarction, secondary to ischemia.
- Treadmill or exercise stress test may provoke chest pain and ECG signs of myocardial ischemia.
- Coronary angiography reveals coronary artery stenosis or obstruction, possible collateral circulation, and the arteries' condition beyond the narrowing.
- Myocardial perfusion imaging with thallium-201, Cardiolite, or Myoview during treadmill exercise detects ischemic areas of the myocardium, visualized as "cold spots."
- Stress echocardiography may show wall motion abnormalities.
- Electron-beam computed tomography identifies calcium within arterial plaque; the more calcium seen, the higher the likelihood of CAD.

Treatment

The goal of treatment in patients with angina is to either reduce myocardial oxygen demand or increase oxygen supply. Therapy consists primarily of nitrates such as nitroglycerin (given sublingually, orally, transdermally, or topically

in ointment form) to dilate coronary arteries and improve blood supply to the heart. Glycoprotein IIb to IIIa inhibitors and antithrombin drugs may be used to reduce the risk of blood clots. BBs may be used to decrease heart rate and lower the heart's oxygen use. CCBs may be used to relax the coronary arteries and all systemic arteries, reducing the heart's workload. ACE inhibitors, diuretics, or other medications may be used to lower blood pressure.

Percutaneous transluminal coronary angioplasty (PTCA) may be performed during cardiac catheterization to compress fatty deposits and relieve occlusion in patients with no calcification and partial occlusion. PTCA carries a certain risk, but the morbidity associated with it is lower than that for surgery. (See *Relieving occlusions with angioplasty*, pages 38 and 39.)

PTCA is an alternative to grafting in elderly patients or others who can't tolerate cardiac surgery. However, patients who have a left main coronary artery occlusion, lesions in extremely tortuous vessels, or occlusions older than 3 months aren't candidates for PTCA.

PTCA can be done along with coronary stenting, or stents may be placed alone. Stents provide a framework to hold an artery open by securing the flaps of the tunica media against an artery wall. Intravascular coronary artery stenting is done to reduce the incidence of restenosis. Prosthetic cylindrical stents made of stainless steel coil are positioned at the site of occlusion. Drug-eluting stents have proven to be safe and effective and have a lower rate of restenosis when compared with bare-metal stents.

Laser angioplasty corrects occlusion by vaporizing fatty deposits with the excimer, or hot-tipped laser device. Percutaneous myocardial revascularization uses a laser to create channels in the heart muscle to improve perfusion to the myocardium. A carbon dioxide laser is used to create transmural channels from the epicardium to the myocardium, extending into the left ventricle. This technique is also known as transmyocardial revascularization and appears to be effective for severe symptoms. In addition, a stent may be placed in the artery to act as a scaffold to hold the artery open. Obstructive lesions may necessitate coronary artery bypass graft (CABG) surgery and the use of vein grafts.

A surgical technique available as an alternative to traditional CABG surgery is minimally invasive coronary artery bypass surgery, also known as *laparoscopic surgery*. This procedure requires a shorter recovery period and has fewer postoperative complications. Instead of sawing open the patient's sternum and spreading the ribs apart, several small cuts are made in the torso through which small surgical instruments and fiber-optic cameras are inserted. This procedure was initially designed to correct blockages in just one or two easily reached arteries; it may not be suitable for more complicated cases.

Coronary brachytherapy, which involves delivering beta or gamma radiation into the coronary arteries, may be used in patients who've undergone stent implantation in a coronary artery but then developed such problems as diffuse in-stent restenosis. Brachytherapy is a promising technique, but its use is restricted to the treatment of stent-related problems because of complications and the unknown long-term effects of the radiation. However, in some facilities, brachytherapy is being studied as a first-line treatment of CAD.

PREVENTION *Because CAD is so widespread, prevention is of great importance. Encourage dietary restrictions aimed at reducing intake of calories (in obesity) and salt, saturated fats, and cholesterol, in order to minimize the risk, especially when supplemented with regular exercise. Also, encourage the patient to stop smoking and to reduce stress. Other preventive actions to encourage include control of hypertension, control of elevated serum cholesterol or triglyceride levels (with antilipemics), and measures to minimize platelet aggregation and the danger of blood clots (with aspirin or other antiplatelet drugs).*

Special considerations

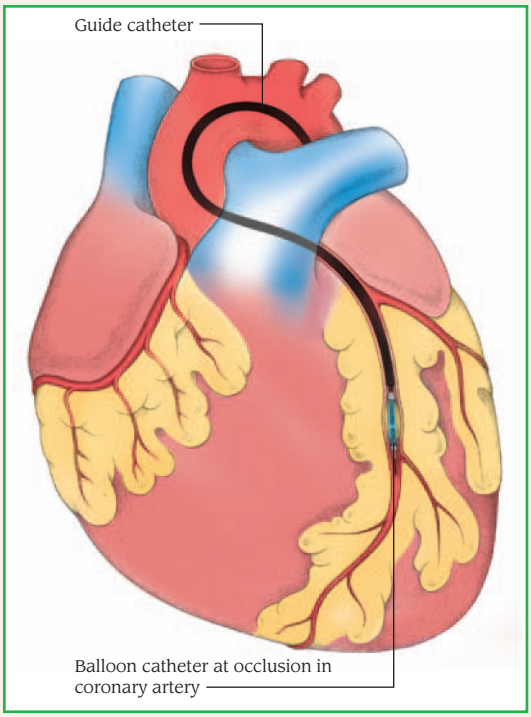
- During anginal episodes, monitor blood pressure and heart rate. Take an ECG during anginal episodes and before administering nitroglycerin or other nitrates. Record duration of pain, amount of medication required to relieve it, and accompanying symptoms.
- Keep nitroglycerin available for immediate use. Instruct the patient to call immediately whenever feeling chest, arm, or neck pain.
- Before cardiac catheterization, explain the procedure to the patient. Make sure the patient knows why it's necessary, understands the risks, and realizes that it may indicate a need for surgery.
- After catheterization, review the expected course of treatment with the patient and family. Monitor the catheter site for bleeding. Also, check for distal pulses. To counter the dye's diuretic effect, make sure the patient drinks plenty of fluids. Assess potassium levels.
- If the patient is scheduled for surgery, explain the procedure to the patient and family. Give them a tour of the ICU and introduce them to the staff.

Relieving occlusions with angioplasty

Percutaneous transluminal coronary angioplasty can open an occluded coronary artery without opening the chest—an important advantage over bypass surgery. First, coronary angiography must confirm the presence and location of the arterial occlusion. Then, the physician threads a guide catheter through the patient's femoral or radial artery into the coronary artery under fluoroscopic guidance, as shown at right.

When angiography shows the guide catheter positioned at the occlusion site, the physician carefully inserts a smaller double-lumen balloon catheter through the guide catheter and directs the balloon through the occlusion (opposite page, left). A marked pressure gradient will be obvious.

The physician alternately inflates and deflates the balloon until an angiogram verifies successful arterial dilation (opposite page, right) and the pressure gradient has decreased.



◆ After surgery, monitor blood pressure, intake and output, breath sounds, chest tube drainage, and ECG, watching for signs of ischemia and arrhythmias. Also, observe for and treat chest pain and possible dye reactions. Give vigorous chest physiotherapy and guide the patient in removal of secretions through deep breathing, coughing, and expectoration of mucus.

◆ Before discharge, stress the need to follow the prescribed drug regimen (e.g., antihypertensives, nitrates, and antilipemics), exercise program, and diet. Encourage regular, moderate exercise. Refer the patient to a self-help program to stop smoking.

MYOCARDIAL INFARCTION

Causes and incidence

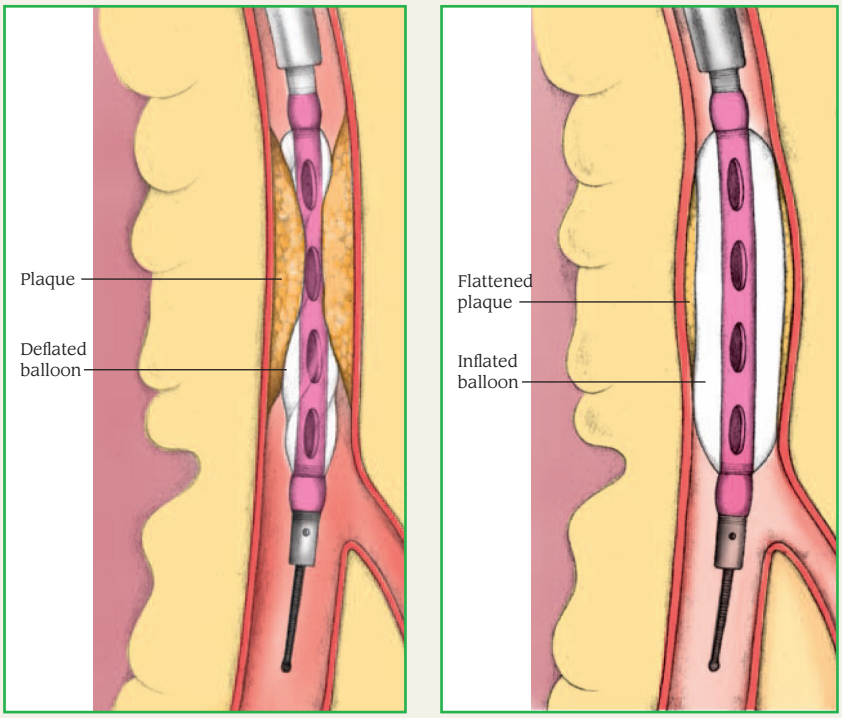
MI, commonly known as a *heart attack* and part of a broader category of disease known as *acute coronary syndrome*, results from prolonged myocardial ischemia due to reduced blood flow through one of the coronary arteries. (See *Tissue destruction in myocardial infarction*, page 40.) In cardiovascular disease, the leading cause of death in the United States and Western Europe, death usually results from the cardiac damage or complications of MI. (See *Complications of myocardial infarction*, page 41.)

Incidence is high: About 1 million patients visit the hospital each year with an MI, and another 120,000 people die from MI-related complications without seeking medical care. Men and postmenopausal women are more susceptible to MI than premenopausal women, although incidence is rising among females, especially those who smoke and take hormonal contraceptives.

Mortality is high when treatment is delayed, and almost one half of sudden deaths due to an MI occur before hospitalization, within 1 hour of the onset of symptoms. The prognosis improves if vigorous treatment begins immediately.

Predisposing risk factors include:

◆ diabetes mellitus
◆ drug use, especially cocaine
◆ elevated serum triglyceride, total cholesterol, and low-density lipoprotein levels
◆ hypertension
◆ obesity or excessive intake of saturated fats, carbohydrates, or salt
◆ positive family history
◆ sedentary lifestyle



- smoking
- stress or a type A personality

Pathophysiology

The site of the MI depends on the vessels involved. Occlusion of the circumflex branch of the left coronary artery causes a lateral wall infarction; occlusion of the anterior descending branch of the left coronary artery, an anterior wall infarction. True posterior or inferior wall infarctions generally result from occlusion of the right coronary artery or one of its branches. Right ventricular infarctions can also result from right coronary artery occlusion, can accompany inferior infarctions, and may cause right-sided heart failure.

Signs and symptoms

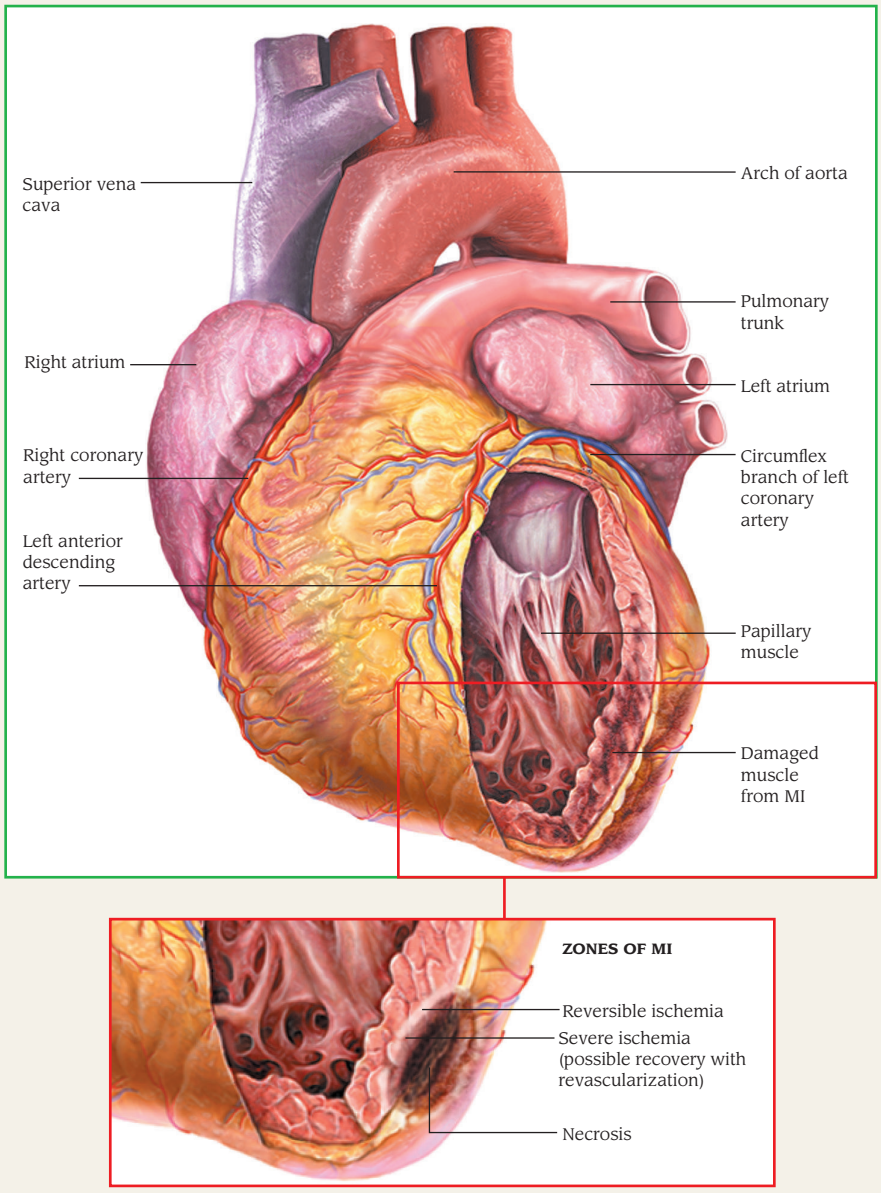
The cardinal symptom of MI is persistent, crushing substernal pain that may radiate to the left arm, jaw, neck, or shoulder blades. Such pain is usually described as heavy, squeezing, or crushing, and may persist for 12 hours or more. However, in some MI patients—particularly elderly people or those with diabetes—pain may not occur at all; in others, it may be mild and confused with indigestion. In patients with CAD, angina of increasing frequency, severity, or duration (especially if not provoked by exertion, a heavy meal, or cold and wind) may signal impending infarction.

Other clinical effects include a feeling of impending doom, fatigue, nausea, vomiting, and shortness of breath. Some patients may have no symptoms. The patient may experience catecholamine responses, such as coolness in extremities, perspiration, anxiety, and restlessness. Fever is unusual at the onset of an MI, but a low-grade temperature elevation may develop during the next few days. Blood pressure varies; hypotension or hypertension may be present.

The most common post-MI complications include recurrent or persistent chest pain, arrhythmias, left-sided heart failure (resulting in heart failure or acute pulmonary edema), and cardiogenic shock. Unusual but potentially lethal complications that may develop soon after infarction include thromboembolism; papillary muscle dysfunction or rupture, causing mitral insufficiency; rupture of the ventricular septum, causing VSD; rupture of the myocardium; and ventricular aneurysm. Up to several months after infarction, Dressler syndrome (pericarditis, pericardial

Tissue destruction in myocardial infarction

A myocardial infarction results from prolonged myocardial ischemia due to reduced blood flow through one or more of the coronary arteries.



Complications of myocardial infarction

Complication	*Diagnosis*	*Treatment*
Arrhythmias	♦ Electrocardiogram (ECG) shows premature ventricular contractions, ventricular tachycardia, or ventricular fibrillation; in inferior wall myocardial infarction (MI), bradycardia and junctional rhythms or atrioventricular block; in anterior wall MI, tachycardia or heart block	♦ Antiarrhythmics, atropine, and pacemaker; cardioversion for tachycardia
Heart failure	♦ In left-sided heart failure, chest X-rays show venous congestion, cardiomegaly, and Kerley B lines ♦ Catheterization shows increased pulmonary artery pressure (PAP) and central venous pressure	♦ Diuretics, angiotensin-converting enzyme inhibitors, vasodilators, inotropic agents, cardiac glycosides, and beta-adrenergic blockers
Cardiogenic shock	♦ Catheterization shows decreased cardiac output and increased PAP and pulmonary artery wedge pressure (PAWP) ♦ Signs include hypertension, tachycardia, S_3, S_4, decreased levels of consciousness, decreased urine output, jugular vein distention, and cool, pale skin	♦ I.V. fluids, vasodilators, diuretics, cardiac glycosides, intra-aortic balloon pump (IABP), and beta-adrenergic stimulants
Rupture of left ventricular papillary muscle	♦ Auscultation reveals an apical holosystolic murmur. Inspection of jugular vein pulse or hemodynamic monitoring shows increased v waves ♦ Dyspnea is prominent ♦ Color-flow and Doppler echocardiogram show mitral insufficiency. Pulmonary artery catheterization shows increased PAP and PAWP	♦ Nitroprusside (Nitropress) ♦ IABP ♦ Surgical replacement of the mitral valve with possible concomitant myocardial revascularization (in patients with significant coronary artery disease)
Ventricular septal rupture	♦ In left-to-right shunt, auscultation reveals a holosystolic murmur and thrill ♦ Catheterization shows increased PAP and PAWP ♦ Confirmation is by increased oxygen saturation of the right ventricle and pulmonary artery	♦ Surgical correction, IABP, nitroglycerin, nitroprusside, low-dose inotropic agents, or pacemaker
Pericarditis or Dressler syndrome	♦ Auscultation reveals a friction rub ♦ Chest pain is relieved by sitting up	♦ Aspirin or NSAIDs
	♦ Chest X-ray may show cardiomegaly ♦ ECG may show arrhythmias and persistent ST-segment elevation ♦ Left ventriculography shows altered or paradoxical left ventricular motion	♦ Cardioversion, defibrillation, antiarrhythmics, vasodilators, anticoagulants, cardiac glycosides, and diuretics (if conservative treatment fails, surgical resection is necessary)
Thromboembolism	♦ Severe dyspnea and chest pain or neurologic changes ♦ Nuclear scan shows ventilation–perfusion mismatch ♦ Angiography shows arterial blockage	♦ Oxygen and heparin