

# Cardiovascular Disorders

## Introduction

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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.



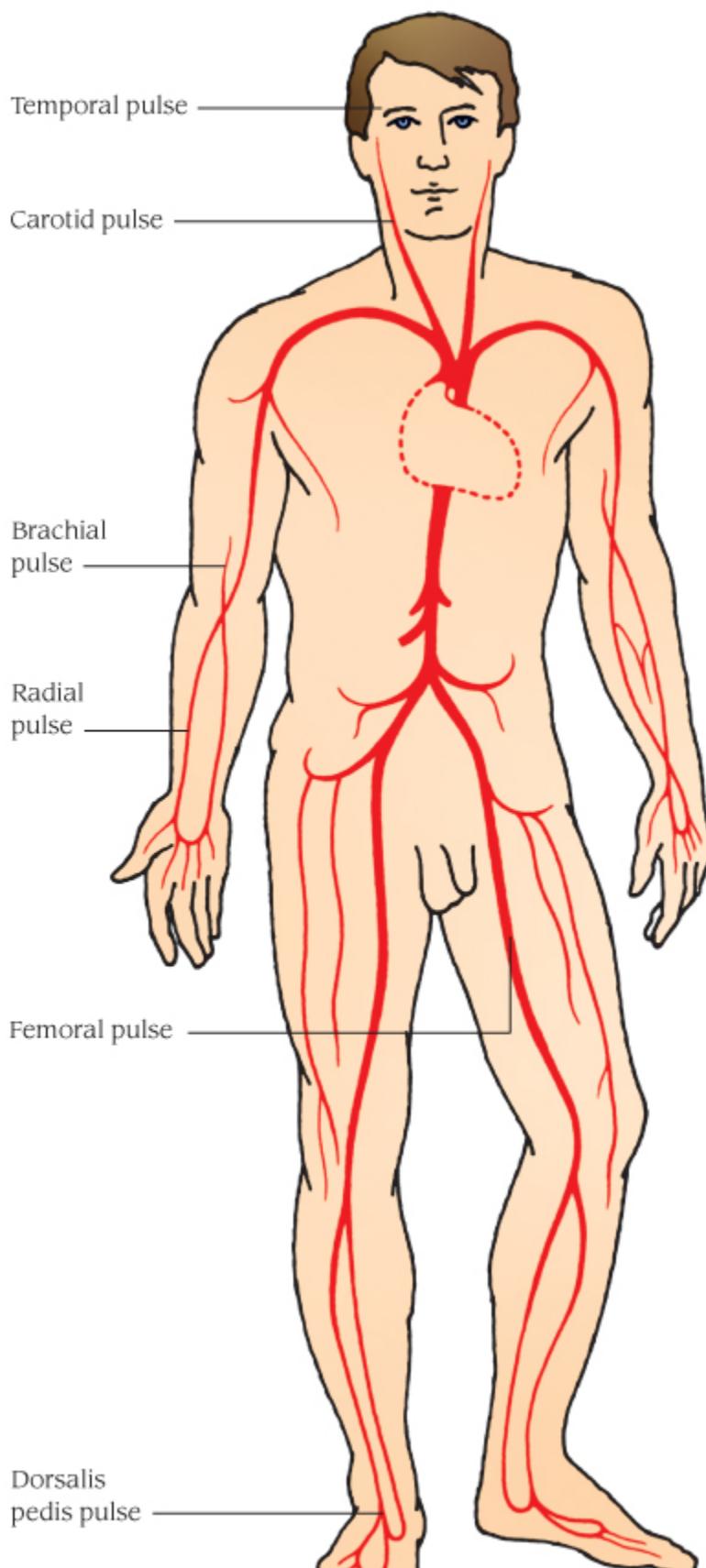
**ELDER TIP** Aging contributes to arterial and venous insufficiency as the strength and elasticity of blood vessels decrease.

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

## 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 ( $\geq 15$  mm Hg decrease in systolic blood pressure during inspiration), often seen in cardiac tamponade.





- ◆ *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

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*.)

### 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.

- ◆ 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 ( $\frac{3}{8}$ " to  $\frac{3}{4}$ " [1 to 2 cm]) and position (in the mitral area)—possible signs of left ventricular hypertrophy, left-sided valvular disease, or right ventricular disease.
- ◆ 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<sub>1</sub> and S<sub>2</sub> (systolic), between S<sub>2</sub> and the following S<sub>1</sub> (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.)

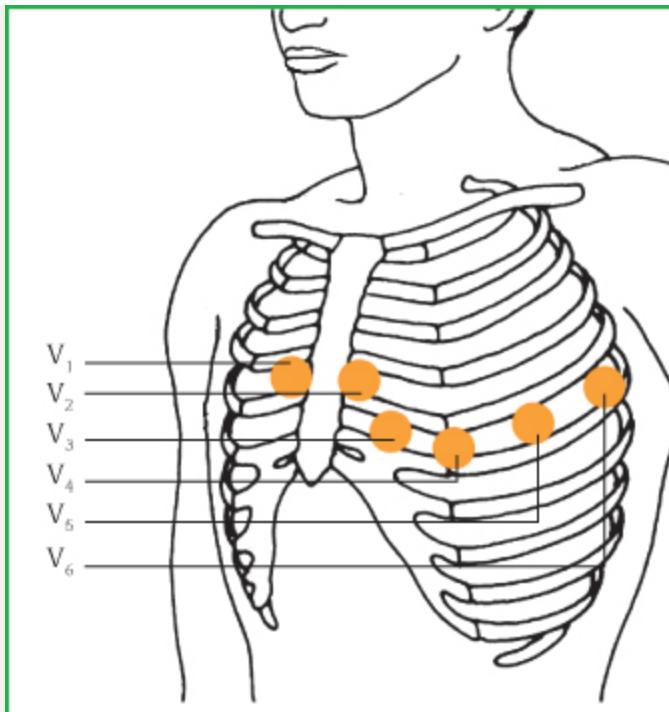
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## Positioning chest electrodes

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



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 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—creatinine 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**

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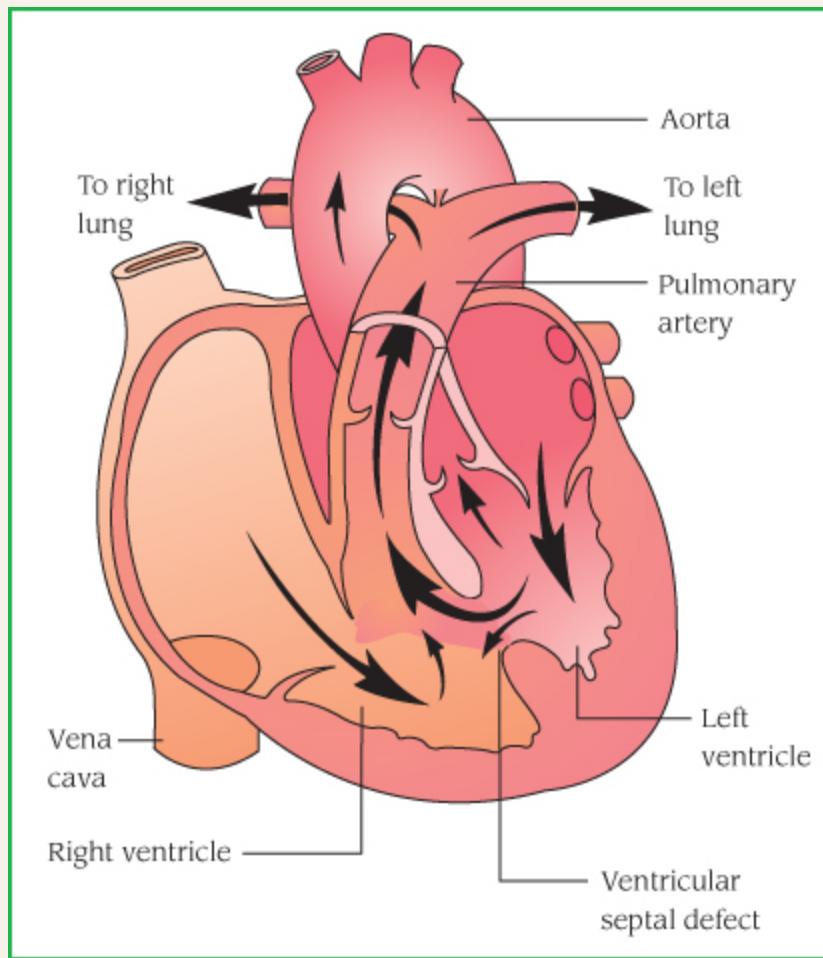
### **VENTRICULAR SEPTAL DEFECT**

#### **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.)

## 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.



## 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 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<sub>2</sub> 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<sub>2</sub> 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*

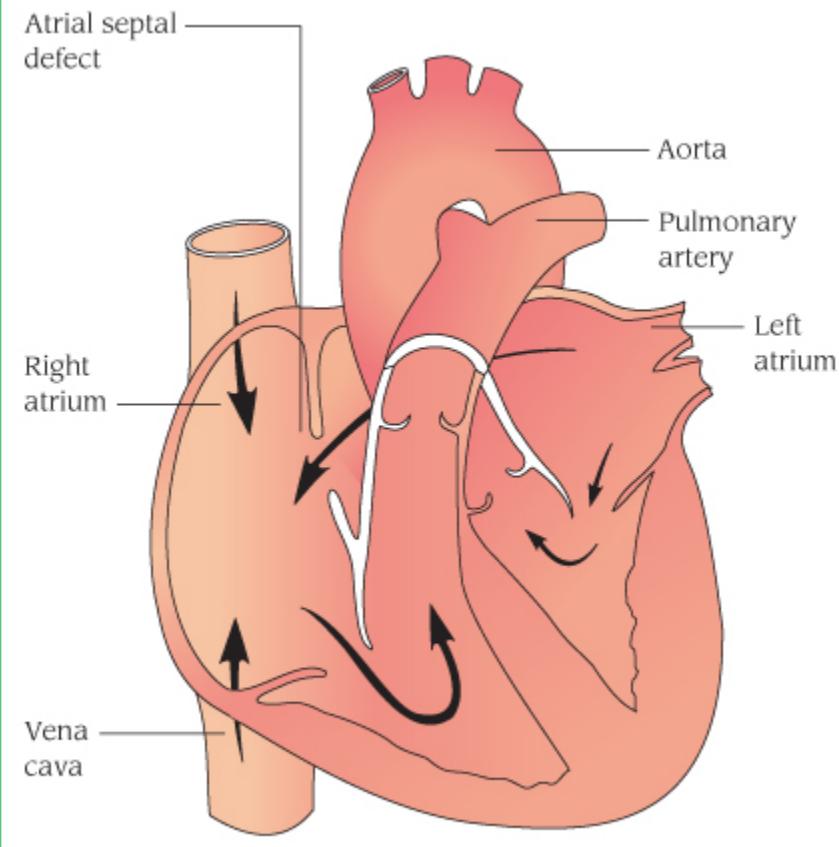
occurs in the superior–posterior portion of the atrial septum, sometimes extending into the vena cava, and is almost always associated with abnormal drainage of pulmonary veins into the right atrium; *ostium primum defect* occurs in the inferior portion of the septum primum and is usually associated 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*.)

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## 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<sub>2</sub>, 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<sub>2</sub>. A pulmonary ejection click and an audible S<sub>4</sub> 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

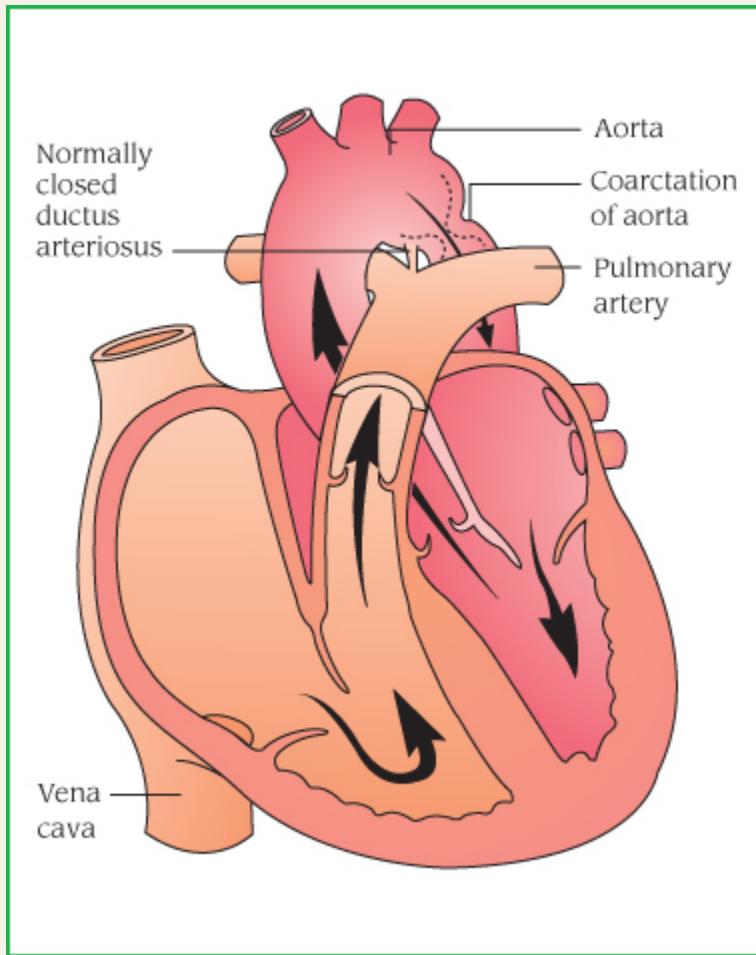
### Causes and Incidence

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*.)

### 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.



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 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<sub>2</sub>, and an S<sub>4</sub>.

## 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

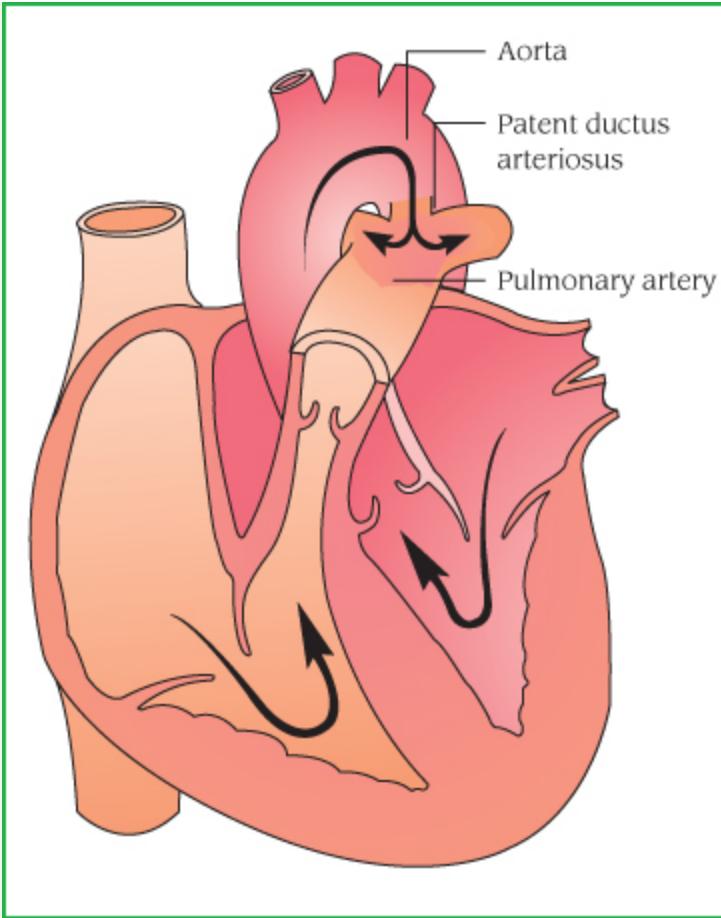
The ductus arteriosus is a fetal blood vessel that connects the pulmonary artery to the descending aorta. In PDA, the lumen of the ductus remains open after birth. This creates a left-to-right shunt of blood from the aorta to the pulmonary artery and results in recirculation of arterial blood through 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<sub>2</sub>. 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

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### 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<sub>2</sub>. 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**

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### **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<sub>3</sub> and S<sub>4</sub> gallops, a faint S<sub>1</sub>, 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.

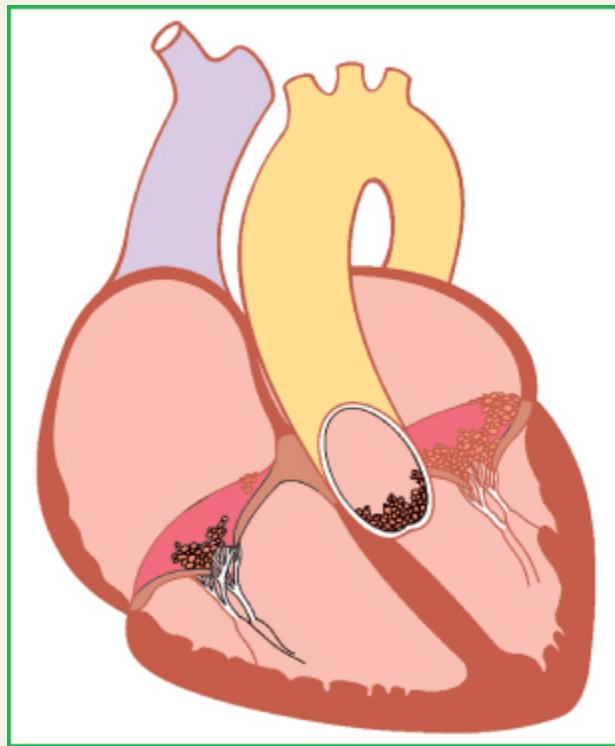
### Pathophysiology

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.

## Degenerative Changes in Endocarditis

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



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 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*.)



## 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.

## 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*.)

## Patterns of cardiac pain

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

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



**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. 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

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### 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*.

## Types of valvular heart disease

| <b>Causes and incidence</b>   | <b>Signs and symptoms</b>  | <b>Diagnostic measures</b>  |
|---|--|---|
| <b>Aortic insufficiency</b>   |  |   |
| <ul style="list-style-type: none"> <li>◆ Results from rheumatic fever, syphilis, hypertension, endocarditis, or may be idiopathic</li> <li>◆ Associated with Marfan syndrome</li> <li>◆ Most common in males</li> <li>◆ Associated with ventricular septal defect, even after surgical closure</li> </ul> | <ul style="list-style-type: none"> <li>◆ Dyspnea, cough, fatigue, palpitations, angina, syncope</li> <li>◆ Pulmonary venous congestion, heart failure, pulmonary edema (left-sided heart failure), “pulsating” nail beds</li> <li>◆ Rapidly rising and collapsing pulses (pulsus bisferiens), cardiac arrhythmias, wide pulse pressure in severe insufficiency</li> <li>◆ Auscultation: reveals S<sub>3</sub> and diastolic blowing murmur at left sternal border</li> <li>◆ Palpation and visualization of apical impulse in chronic disease</li> </ul> | <ul style="list-style-type: none"> <li>◆ Cardiac catheterization: reduction in arterial diastolic pressures, aortic insufficiency, other valvular abnormalities, and increased left ventricular end-diastolic pressure</li> <li>◆ X-ray: left ventricular enlargement, pulmonary vein congestion</li> <li>◆ Echocardiography: left ventricular enlargement, alterations in mitral valve movement (indirect indication of aortic valve disease), and mitral thickening</li> <li>◆ Electrocardiography (ECG): sinus tachycardia, left ventricular hypertrophy, and left atrial hypertrophy in severe disease</li> </ul> |
| <b>Aortic stenosis</b>  |  |   |

| <b>Causes and incidence</b>  | <b>Signs and symptoms</b>  | <b>Diagnostic measures</b>  |
|--|--|---|
| <ul style="list-style-type: none"> <li>◆ Results from congenital aortic bicuspid valve (associated with coarctation of the aorta), congenital stenosis of valve cusps, rheumatic fever, or atherosclerosis in elderly persons</li> <li>◆ Most common in males</li> </ul> | <ul style="list-style-type: none"> <li>◆ Dyspnea on exertion, paroxysmal nocturnal dyspnea, fatigue, syncope, angina, palpitations</li> <li>◆ Pulmonary venous congestion, heart failure, pulmonary edema</li> <li>◆ Diminished carotid pulses, decreased cardiac output, cardiac arrhythmias; may have pulsus alternans</li> <li>◆ Auscultation: reveals systolic murmur at base or in carotids and, possibly, S<sub>4</sub></li> </ul> | <ul style="list-style-type: none"> <li>◆ Cardiac catheterization: pressure gradient across valve (indicating obstruction), increased left ventricular end-diastolic pressures</li> <li>◆ X-ray: valvular calcification, left ventricular enlargement, and pulmonary venous congestion</li> <li>◆ Echocardiography: thickened aortic valve and left ventricular wall</li> <li>◆ ECG: left ventricular hypertrophy</li> </ul> |

### Mitral insufficiency

| <b>Causes and incidence</b>  | <b>Signs and symptoms</b>  | <b>Diagnostic measures</b>   |
|--|--|--|
| <ul style="list-style-type: none"> <li>◆ Results from rheumatic fever, hypertrophic cardiomyopathy, mitral valve prolapse, myocardial infarction, severe left-sided heart failure, or ruptured chordae tendineae</li> <li>◆ Associated with other congenital anomalies such as transposition of the great arteries</li> <li>◆ Rare in children without other congenital anomalies</li> </ul> | <ul style="list-style-type: none"> <li>◆ Orthopnea, dyspnea, fatigue, angina, palpitations</li> <li>◆ Peripheral edema, jugular vein distention (JVD), hepatomegaly (right-sided heart failure)</li> <li>◆ Tachycardia, crackles, pulmonary edema</li> <li>◆ Auscultation: reveals holosystolic murmur at apex, possible split S<sub>2</sub>, and S<sub>3</sub></li> </ul> | <ul style="list-style-type: none"> <li>◆ 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</li> <li>◆ X-ray: left atrial and ventricular enlargement, pulmonary venous congestion</li> <li>◆ Echocardiography: abnormal valve leaflet motion, left atrial enlargement</li> <li>◆ ECG: left atrial and ventricular hypertrophy, sinus tachycardia, and atrial fibrillation</li> </ul> |

### **Mitral stenosis**

| <b>Causes and incidence</b>   | <b>Signs and symptoms</b>  | <b>Diagnostic measures</b>  |
|---|--|---|
| <ul style="list-style-type: none"> <li>◆ Results from rheumatic fever (most common cause)</li> <li>◆ Most common in females</li> <li>◆ May be associated with other congenital anomalies</li> </ul> | <ul style="list-style-type: none"> <li>◆ Dyspnea on exertion, paroxysmal nocturnal dyspnea, orthopnea, weakness, fatigue, palpitations</li> <li>◆ Peripheral edema, JVD, ascites, hepatomegaly (right-sided heart failure in severe pulmonary hypertension)</li> <li>◆ Crackles, cardiac arrhythmias (atrial fibrillation), signs of systemic emboli</li> <li>◆ Auscultation: reveals loud S<sub>1</sub> or opening snap and diastolic murmur at apex</li> </ul> | <ul style="list-style-type: none"> <li>◆ Cardiac catheterization: diastolic pressure gradient across valve; elevated left atrial pressure and PAWP (&gt;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</li> <li>◆ X-ray: left atrial and ventricular enlargement, enlarged pulmonary arteries, and mitral valve calcification</li> <li>◆ Echocardiography: thickened mitral valve leaflets, left atrial enlargement</li> <li>◆ ECG: left atrial hypertrophy, atrial fibrillation, right ventricular hypertrophy, and right axis deviation</li> </ul> |

### Mitral valve prolapse syndrome

| <b>Causes and incidence</b>   | <b>Signs and symptoms</b>  | <b>Diagnostic measures</b>  |
|---|--|---|
| <ul style="list-style-type: none"> <li>◆ Can be genetic or associated with conditions such as Ehlers–Danlos syndrome, Marfan syndrome, Graves disease, and muscular dystrophy</li> <li>◆ Most commonly affects young women but may occur in both sexes and in all age groups</li> </ul> | <ul style="list-style-type: none"> <li>◆ May produce no signs</li> <li>◆ Chest pain, palpitations, headache, fatigue, exercise intolerance, dyspnea, light-headedness, syncope, mood swings, anxiety, panic attacks</li> <li>◆ Auscultation: typically reveals mobile, midsystolic click, with or without mid-to-late systolic murmur</li> </ul> | <ul style="list-style-type: none"> <li>◆ Two-dimensional echocardiography: prolapse of mitral valve leaflets into left atrium</li> <li>◆ Color-flow Doppler studies: mitral insufficiency</li> <li>◆ Resting ECG: ST-segment changes, biphasic or inverted T waves in leads II, III, or AV</li> <li>◆ Exercise ECG: evaluates chest pain and arrhythmias</li> </ul> |
| <b>Pulmonic insufficiency</b>   |  |   |
| <ul style="list-style-type: none"> <li>◆ May be congenital or may result from pulmonary hypertension</li> <li>◆ May rarely result from prolonged use of pressure-monitoring catheter in the pulmonary artery</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Dyspnea, weakness, fatigue, chest pain</li> <li>◆ Peripheral edema, JVD, hepatomegaly (right-sided heart failure)</li> <li>◆ Auscultation: reveals diastolic murmur in pulmonic area</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Cardiac catheterization: pulmonic insufficiency, increased right ventricular pressure, and associated cardiac defects</li> <li>◆ X-ray: right ventricular and pulmonary arterial enlargement</li> <li>◆ ECG: right ventricular or right atrial enlargement</li> </ul>  |
| <b>Pulmonic stenosis</b>  |  |   |

| <b>Causes and incidence</b>   | <b>Signs and symptoms</b>  | <b>Diagnostic measures</b>   |
|---|--|--|
| <ul style="list-style-type: none"> <li>◆ Results from congenital stenosis of valve cusp or rheumatic heart disease (infrequent)</li> <li>◆ Associated with other congenital heart defects such as tetralogy of Fallot</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Asymptomatic or symptomatic with dyspnea on exertion, fatigue, chest pain, syncope</li> <li>◆ May lead to peripheral edema, JVD, hepatomegaly (right-sided heart failure)</li> <li>◆ Auscultation: reveals systolic murmur at left sternal border, split S<sub>2</sub> with delayed or absent pulmonic component</li> </ul> | <ul style="list-style-type: none"> <li>◆ Cardiac catheterization: increased right ventricular pressure, decreased PAP, and abnormal valve orifice</li> <li>◆ ECG: may show right ventricular hypertrophy, right axis deviation, right atrial hypertrophy, and atrial fibrillation</li> </ul>   |
| <b>Tricuspid insufficiency</b>  |  |  |
| <ul style="list-style-type: none"> <li>◆ Results from right-sided heart failure, rheumatic fever and, rarely, trauma and endocarditis</li> <li>◆ Associated with congenital disorders</li> <li>◆ Associated with I.V. drug abuse and infective endocarditis manifesting as tricuspid valve disease</li> </ul> | <ul style="list-style-type: none"> <li>◆ Dyspnea and fatigue</li> <li>◆ May lead to peripheral edema, JVD, hepatomegaly, and ascites (right-sided heart failure)</li> <li>◆ Auscultation: reveals possible S<sub>3</sub> and systolic murmur at lower left sternal border that increases with inspiration</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Right-sided heart catheterization: high atrial pressure, tricuspid insufficiency, decreased or normal cardiac output</li> <li>◆ X-ray: right atrial dilation, right ventricular enlargement</li> <li>◆ Echocardiography: shows systolic prolapse of tricuspid valve, right atrial enlargement</li> <li>◆ ECG: right atrial or right ventricular hypertrophy, atrial fibrillation</li> </ul> |
| <b>Tricuspid stenosis</b>   |  |  |

| <b>Causes and incidence</b>   | <b>Signs and symptoms</b>   | <b>Diagnostic measures</b>  |
|---|---|---|
| <ul style="list-style-type: none"> <li>◆ Results from rheumatic fever</li> <li>◆ May be congenital</li> <li>◆ Associated with mitral or aortic valve disease</li> <li>◆ Most common in women</li> </ul> | <ul style="list-style-type: none"> <li>◆ May be symptomatic with dyspnea, fatigue, syncope</li> <li>◆ Possibly peripheral edema, JVD, hepatomegaly, and ascites (right-sided heart failure)</li> <li>◆ Auscultation: reveals diastolic murmur at lower left sternal border that increases with inspiration</li> </ul> | <ul style="list-style-type: none"> <li>◆ Cardiac catheterization: increased pressure gradient across valve, increased right atrial pressure, decreased cardiac output</li> <li>◆ X-ray: right atrial enlargement</li> <li>◆ Echocardiography: leaflet abnormality, right atrial enlargement</li> <li>◆ ECG: right atrial hypertrophy, right or left ventricular hypertrophy, and atrial fibrillation</li> </ul> |

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

# **Degenerative Cardiovascular Disorders**

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## **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.

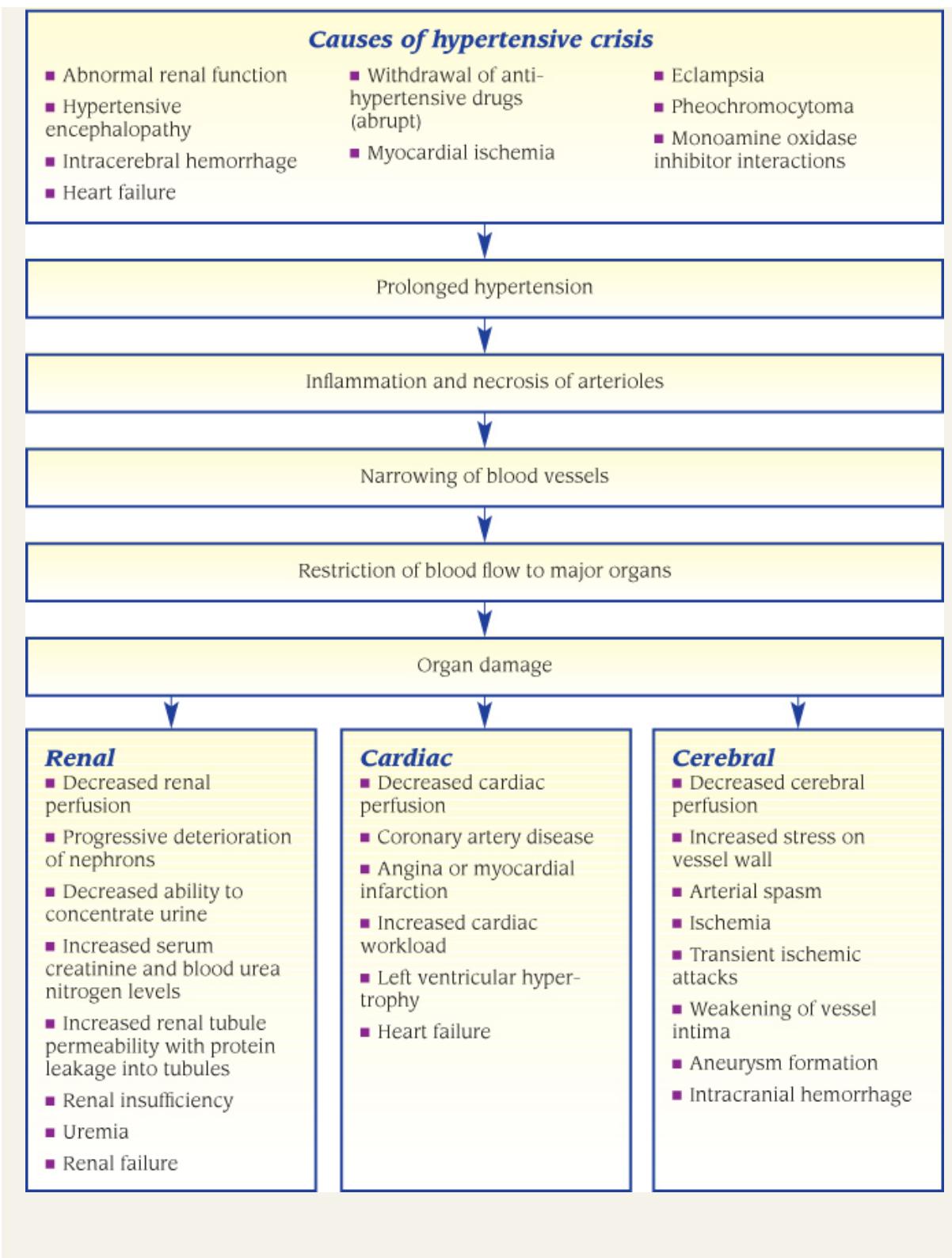
The prognosis is good if this disorder is detected early and treatment begins before complications develop. Severely elevated blood pressure (hypertensive crisis) may be fatal. (See *What happens in a hypertensive crisis*, page 31.)



## **PATHOPHYSIOLOGY**

### **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.



## 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. 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.

## 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.)

## 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.

| <b>Category</b>     | <b>Systolic (mm Hg)</b> |        | <b>Diastolic (mm Hg)</b> |
|---------------------|-------------------------|--------|--------------------------|
| 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                     |

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 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.)



## **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.

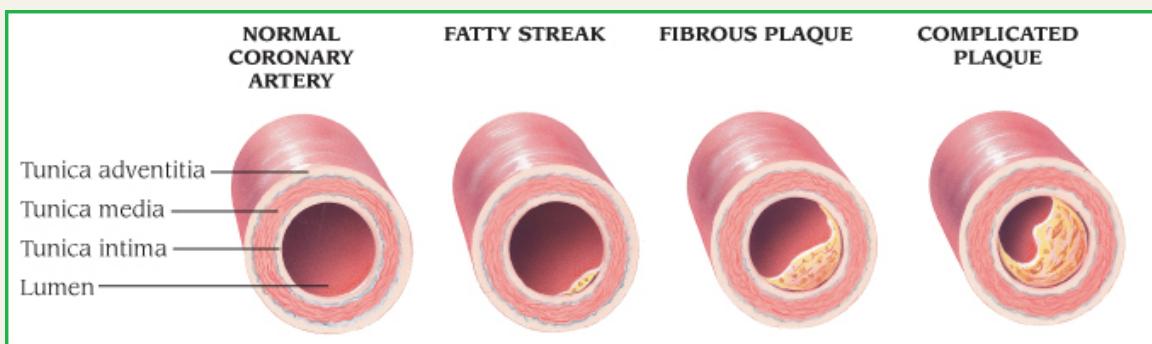
# CORONARY ARTERY DISEASE

## 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.*)

## 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.



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.)

## 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.

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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 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.)

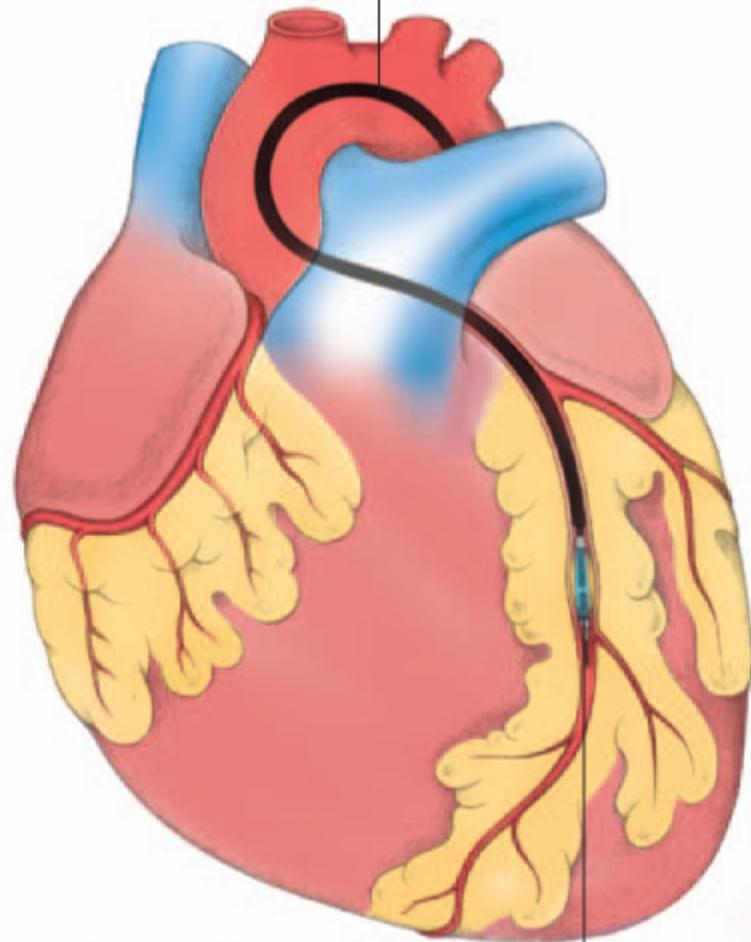
## Relieving Occlusions With Angioplasty

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.

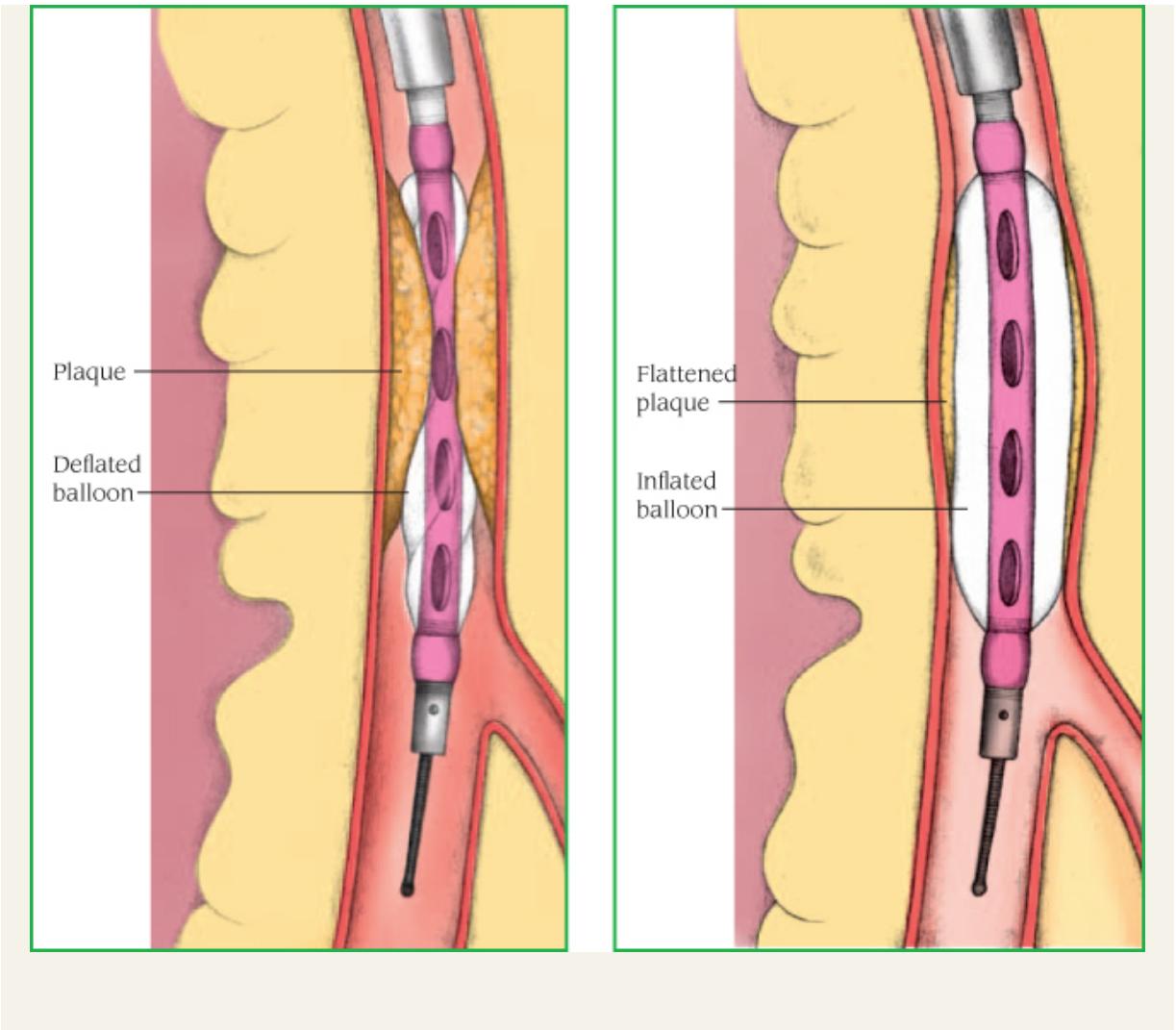
When angiography shows the guide catheter positioned at the occlusion site, the physician carefully inserts a smaller double-lumen balloon catheter through the guide catheter and directs the balloon through the occlusion (opposite page, left). A marked pressure gradient will be obvious.

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

Guide catheter



Balloon catheter at occlusion in  
coronary artery



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.
- ◆ 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

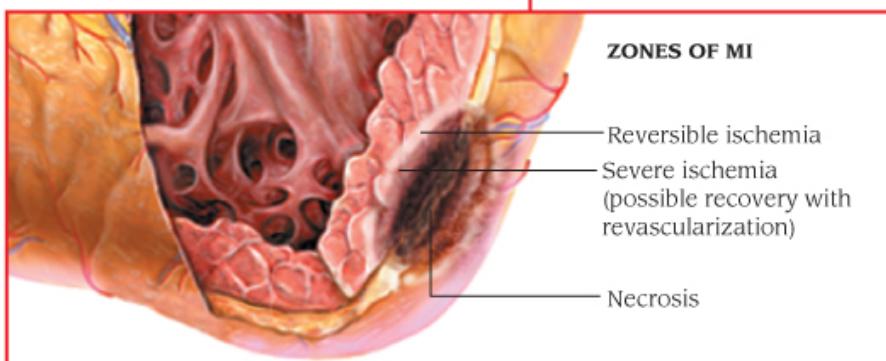
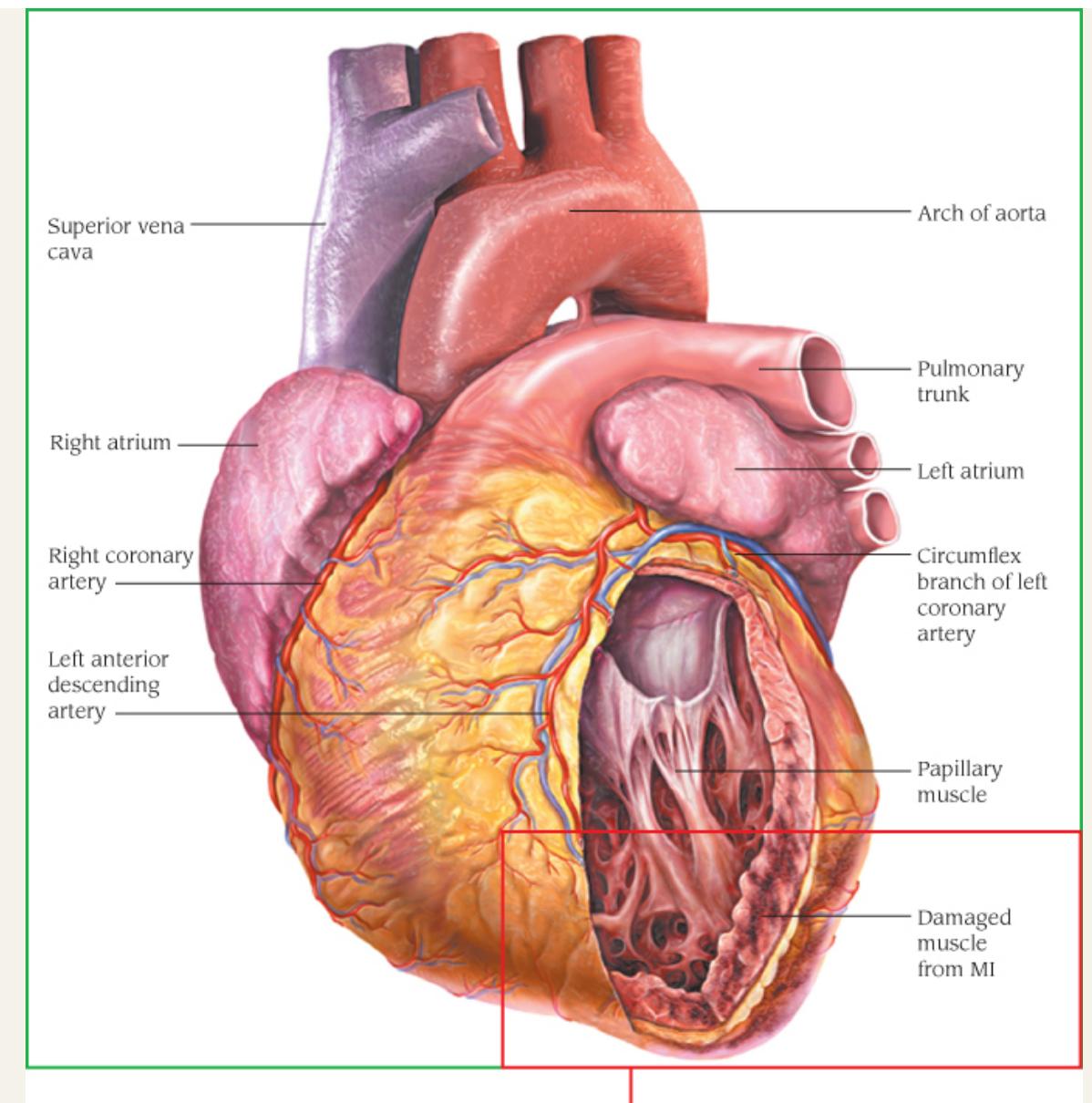
MI, commonly known as a *heart attack* and part of a broader category of disease known as *acute coronary syndrome*, results from prolonged myocardial ischemia due to reduced blood flow through one of the coronary arteries. (See *Tissue destruction in myocardial infarction*, page 40.) In cardiovascular 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.)

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## **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

| <b>Complication</b>                          | <b>Diagnosis</b>   | <b>Treatment</b>   |
|--|--|--|
| Arrhythmias                                  | <ul style="list-style-type: none"> <li>◆ 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</li> </ul>                               | <ul style="list-style-type: none"> <li>◆ Antiarrhythmics, atropine, and pacemaker; cardioversion for tachycardia</li> </ul>  |
| Heart failure                                | <ul style="list-style-type: none"> <li>◆ In left-sided heart failure, chest X-rays show venous congestion, cardiomegaly, and Kerley B lines</li> <li>◆ Catheterization shows increased pulmonary artery pressure (PAP) and central venous pressure</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Diuretics, angiotensin-converting enzyme inhibitors, vasodilators, inotropic agents, cardiac glycosides, and beta-adrenergic blockers</li> </ul>  |
| Cardiogenic shock                            | <ul style="list-style-type: none"> <li>◆ Catheterization shows decreased cardiac output and increased PAP and pulmonary artery wedge pressure (PAWP)</li> <li>◆ Signs include hypertension, tachycardia, S<sub>3</sub>, S<sub>4</sub>, decreased levels of consciousness, decreased urine output, jugular vein distention, and cool, pale skin</li> </ul>            | <ul style="list-style-type: none"> <li>◆ I.V. fluids, vasodilators, diuretics, cardiac glycosides, intra-aortic balloon pump (IABP), and beta-adrenergic stimulants</li> </ul>   |
| Rupture of left ventricular papillary muscle | <ul style="list-style-type: none"> <li>◆ Auscultation reveals an apical holosystolic murmur. Inspection of jugular vein pulse or hemodynamic monitoring shows increased v waves</li> <li>◆ Dyspnea is prominent</li> <li>◆ Color-flow and Doppler echocardiogram show mitral insufficiency. Pulmonary artery catheterization shows increased PAP and PAWP</li> </ul> | <ul style="list-style-type: none"> <li>◆ Nitroprusside (Nitropress)</li> <li>◆ IABP</li> <li>◆ Surgical replacement of the mitral valve with possible concomitant myocardial revascularization (in patients with significant coronary artery disease)</li> </ul> |
| Ventricular septal rupture                   | <ul style="list-style-type: none"> <li>◆ In left-to-right shunt, auscultation reveals a holosystolic murmur and thrill</li> <li>◆ Catheterization shows increased PAP and PAWP</li> <li>◆ Confirmation is by increased oxygen saturation of the right ventricle and pulmonary artery</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Surgical correction, IABP, nitroglycerin, nitroprusside, low-dose inotropic agents, or pacemaker</li> </ul>   |

| <b>Complication</b>               | <b>Diagnosis</b>   | <b>Treatment</b>   |
|-----------------------------------|--|--|
| Pericarditis or Dressler syndrome | <ul style="list-style-type: none"> <li>◆ Auscultation reveals a friction rub</li> <li>◆ Chest pain is relieved by sitting up</li> <li>◆ Chest X-ray may show cardiomegaly</li> <li>◆ ECG may show arrhythmias and persistent ST-segment elevation</li> <li>◆ Left ventriculography shows altered or paradoxical left ventricular motion</li> </ul> | <ul style="list-style-type: none"> <li>◆ Aspirin or NSAIDs</li> </ul>  |
| Thromboembolism                   | <ul style="list-style-type: none"> <li>◆ Severe dyspnea and chest pain or neurologic changes</li> <li>◆ Nuclear scan shows ventilation–perfusion mismatch</li> <li>◆ Angiography shows arterial blockage</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Cardioversion, defibrillation, antiarrhythmics, vasodilators, anticoagulants, cardiac glycosides, and diuretics (if conservative treatment fails, surgical resection is necessary)</li> <li>◆ Oxygen and heparin</li> </ul> |

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 friction rub, chest

pain, fever, leukocytosis and, possibly, pleurisy or pneumonitis) may develop.

## Diagnosis

 **CONFIRMING DIAGNOSIS** *Persistent chest pain, elevated ST segment on ECG, and elevated total CK and CK-MB levels over a 72-hour period are consistent with ST-elevation MI (STEMI). Troponin T or troponin I is also used in the diagnosis because both are specific to cardiac necrosis, and levels rise 6 to 8 hours after onset of ischemia. These labs are also useful when no ST-segment elevation occurs, as in non-ST elevation MI (NSTEMI).*

Auscultation may reveal diminished heart sounds, gallops, and, in papillary dysfunction, the apical systolic murmur of mitral insufficiency over the mitral valve area.

When clinical features are equivocal, assume that the patient had an MI until tests rule it out. Diagnostic laboratory results include:

- ◆ serial 12-lead ECG—ECG abnormalities may be absent or inconclusive during the first few hours after an MI. When present, characteristic abnormalities include serial ST-segment depression NSTEMI and STEMI.
- ◆ serial serum enzyme levels—CK levels are elevated, specifically, the CK-MB isoenzyme.
- ◆ echocardiography—may show ventricular wall motion abnormalities.
- ◆ nuclear ventriculography scans (MUGA or radionuclide ventriculography)—using I.V. radioactive substance, can identify acutely damaged muscle by picking up radioactive nucleotide, which appears as a “hot spot” on the film; useful in localizing a recent MI.

## Treatment

The goals of treatment are to relieve chest pain, stabilize heart rhythm, reduce cardiac workload, revascularize the coronary artery, and preserve myocardial tissue. Arrhythmias, the predominant problem during the first 48 hours after the infarction, may require antiarrhythmics, possibly a pacemaker, and, rarely, cardioversion. Arrhythmias are best detected using a 12-lead ECG.

To preserve myocardial tissue is primary percutaneous coronary intervention (PCI) (mechanical reperfusion). PCI has been shown in many studies to be superior to fibrinolysis in the combined end points of death, stroke, and reinfarction when it is performed within 90 minutes of patient arrival. When primary PCI cannot be performed within 90 minutes, thrombolysis is the treatment of choice in STEMI. (See *Comparing thrombolytics*, page 43.)

## Comparing Thrombolytics

If your patient has suffered a myocardial infarction (MI), you must intervene promptly to minimize cardiac damage and avert death. If appropriate, prepare the patient for thrombolytic therapy as ordered.

Thrombolytic drugs enhance the body's natural ability to dispose of blood clots. To lyse (dissolve) fibrin, the essential component of a clot, tissue activators convert plasminogen to plasmin. A nonspecific protease, plasmin, degrades fibrin, fibrinogen, and procoagulant factors (such as factors V, VII, and XII).

Candidates for thrombolytic therapy include patients with acute ST-segment elevation and chest pain that has lasted no more than 6 hours and who have no access to cardiac catheterization. Timely use of thrombolytic agents can restore myocardial perfusion and prevent further injury. When effective, thrombolytic agents relieve chest pain, restore the ST segment to baseline, and induce reperfusion arrhythmias within 30 to 45 minutes.

Contraindications to thrombolytic therapy include surgery within the past 2 months, active bleeding, a history of stroke, intracranial neoplasm, arteriovenous malformation, aneurysm, or uncontrolled hypertension.

Here's how selected thrombolytics open occluded coronary arteries in patients with an acute MI.

### Alteplase (Activase)

This naturally occurring enzyme has been cloned and produced as a drug, alteplase (tissue plasminogen activator). Binding to plasminogen, it catalyzes the conversion of plasminogen to plasmin in the presence of

fibrin. Because of its strong affinity for fibrin, alteplase concentrates at the clot site, resulting in a minimal decrease in the fibrinogen level.

This thrombolytic has a half-life of 5 minutes, so maintaining coronary artery patency depends on continued anticoagulation with heparin. Alteplase doesn't induce antigenic responses; doses may be repeated at any time.

### **Reteplase (Retavase)**

Reteplase, recombinant plasminogen activator, has a half-life of 13 to 16 minutes. Its longer half-life allows it to be administered as a bolus. Two boluses are required.

### **Streptokinase (Streptase)**

Streptokinase, a thrombolytic, is a bacterial protein that binds to circulating plasminogen and catalyzes plasmin formation. Its low specificity for fibrin induces a systemic lytic state and increases the risk of bleeding.

The half-life is ~20 minutes. Like anistreplase, streptokinase is antigenic.

### **Tenecteplase (Tnkase)**

Tenecteplase is a modified form of human tissue plasminogen activator that binds to fibrin and converts plasminogen to plasmin. It's given as a single bolus dose.

### **Urokinase (Abbokinase)**

Naturally produced by the human kidney, urokinase promotes thrombolysis by directly activating the conversion of plasminogen to plasmin.

With a serum half-life of 10 to 20 minutes, urokinase is rapidly cleared by the kidneys and liver. Unlike streptokinase, it doesn't induce an antigenic response. Urokinase isn't given through a peripheral I.V. line to treat an acute MI, but patients who undergo cardiac catheterization may receive it directly in a coronary artery.

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Other treatments consist of:

- ◆ lidocaine, vasopressin, or amiodarone for ventricular arrhythmias, or other drugs, such as procainamide, quinidine, or disopyramide
- ◆ antiplatelet therapy with glycoprotein IIb to IIIa inhibitors, such as clopidogrel for non-STEMI
- ◆ atropine I.V. or a temporary pacemaker for heart block or bradycardia
- ◆ nitroglycerin (sublingual, topical, transdermal, or I.V.); CCBs, such as nifedipine, verapamil, or diltiazem (sublingual, oral, or I.V.); or isosorbide dinitrate (sublingual, oral, or I.V.) to relieve pain by redistributing blood to ischemic areas of the myocardium, increasing cardiac output, and reducing myocardial workload
- ◆ heparin I.V. (usually follows thrombolytic therapy)
- ◆ morphine I.V. for pain and sedation
- ◆ bed rest with bedside commode to decrease cardiac workload
- ◆ oxygen administration at a modest flow rate for 2 to 3 hours (a lower concentration is necessary if the patient has chronic obstructive pulmonary disease)
- ◆ ACE inhibitors for patients with large anterior wall MIs and for those with an MI and a left ventricular ejection fraction less than 40%
- ◆ drugs to increase myocardial contractility or blood pressure
- ◆ BBs, such as carvedilol or atenolol, after acute MI to help prevent reinfarction by reducing the heart's workload
- ◆ aspirin to inhibit platelet aggregation (should be initiated immediately and continued for years)
- ◆ pulmonary artery catheterization to detect left- or right-sided heart failure and to monitor the patient's response to treatment.

## Special Considerations

Care for patients who have suffered an MI is directed toward detecting complications; preventing further myocardial damage; and promoting comfort, rest, and emotional well-being. Most MI patients receive treatment in the ICU or on a telemetry unit, where they're under constant observation for complications.

- ◆ On admission, monitor and record the patient's ECG, blood pressure, temperature, and heart and breath sounds.
- ◆ Assess and record the severity and duration of pain, and administer analgesics. Avoid I.M. injections; absorption from the muscle is

unpredictable, and bleeding is likely if the patient is receiving thrombolytic therapy.

- ◆ Check the patient's blood pressure after giving nitroglycerin, especially the first dose.
- ◆ Frequently monitor the ECG to detect rate changes or arrhythmias. Place rhythm strips in the patient's chart periodically for evaluation.
- ◆ During episodes of chest pain, obtain 12-lead ECG (before and after nitroglycerin therapy as well), blood pressure, and pulmonary artery catheter measurements, and monitor them for changes.
- ◆ Watch for signs and symptoms of fluid retention (crackles, cough, tachypnea, and edema), which may indicate impending heart failure. Carefully monitor daily weight, intake and output, respirations, serum enzyme levels, and blood pressure. Auscultate for adventitious breath sounds periodically (patients on bed rest frequently have atelectatic crackles, which disappear after coughing), for S<sub>3</sub> or S<sub>4</sub> gallops, and for new-onset heart murmurs.
- ◆ Organize patient care and activities to maximize periods of uninterrupted rest.
- ◆ Initiate a cardiac rehabilitation program. This usually includes education regarding heart disease, exercise, and emotional support for the patient and family.
- ◆ Ask the dietary department to provide a clear liquid diet until nausea subsides. A low-cholesterol, low-sodium, low-fat, high-fiber diet may be prescribed.
- ◆ Provide a stool softener to prevent straining during defecation, because this causes vagal stimulation and may slow the heart rate. Allow use of a bedside commode and provide as much privacy as possible.
- ◆ Assist with range-of-motion exercises. If the patient is completely immobilized by a severe MI, turn often. Antiembolism stockings help prevent venostasis and thrombophlebitis.
- ◆ Provide emotional support and help reduce stress and anxiety. Explain procedures and answer questions. Explaining the ICU environment and routine can ease anxiety. Involve the patient's family in the care as much as possible.

To prepare the patient for discharge:

- ◆ Thoroughly explain dosages and therapy to promote compliance with the prescribed medication regimen and other treatment measures. Warn about drug adverse effects, and advise the patient to watch for and report signs of toxicity (anorexia, nausea, vomiting, and yellow vision, e.g., if the patient is receiving digoxin).
- ◆ Review dietary restrictions with the patient. If the patient must follow a low-sodium or low-fat and low-cholesterol diet, provide a list of foods that the patient should avoid. Ask the dietitian to speak to the patient and family.
- ◆ Counsel the patient to resume sexual activity progressively.
- ◆ Advise the patient to report typical or atypical chest pain. Postinfarction syndrome may develop, producing chest pain that must be differentiated from recurrent MI, pulmonary infarct, or heart failure.
- ◆ If the patient has a Holter monitor in place, explain its purpose and use.
- ◆ Stress the need to stop smoking.
- ◆ Encourage participation in a cardiac rehabilitation program.
- ◆ Review follow-up procedures and office visits with the patient.



## PREVENTION

- ◆ *Instruct patient to practice heart-healthy living, with a heart-healthy diet, regular exercise, stress reduction and preventive care, maintenance of a healthy weight, smoking cessation, and abstinence from alcohol and illegal drugs, especially cocaine.*
- ◆ *Suggest a daily aspirin regimen for patients with CAD or history of an MI.*

## HEART FAILURE

### Causes and Incidence

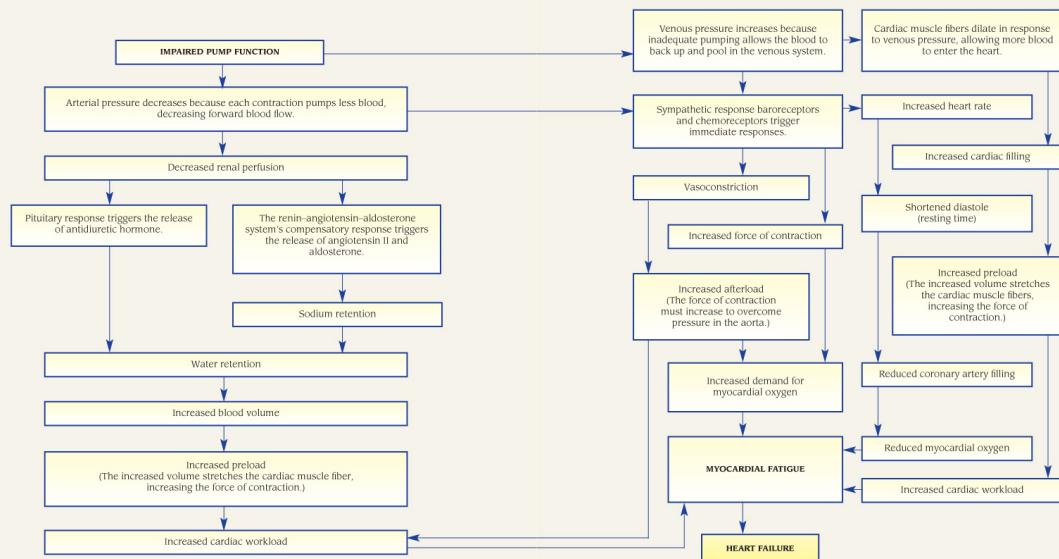
Heart failure is a syndrome characterized by myocardial dysfunction that leads to impaired pump performance (diminished cardiac output) or to frank heart failure and abnormal circulatory congestion. Congestion of systemic venous circulation may result in peripheral edema or hepatomegaly; congestion of pulmonary circulation may cause pulmonary edema, an acute life-threatening emergency. Pump failure usually occurs in a damaged left ventricle (left-sided heart failure) but may occur in the right ventricle (right-

sided heart failure) either as a primary disorder or secondary to left-sided heart failure. Sometimes, left- and right-sided heart failure develop simultaneously. (See *What happens in heart failure*, pages 44 and 45.)



## PATHOPHYSIOLOGY WHAT HAPPENS IN HEART FAILURE

Heart failure occurs when cardiac output is inadequate to meet the body's needs. The pathophysiology of heart failure is shown in the flow chart below.



Heart failure may result from a primary abnormality of the heart muscle such as an infarction, inadequate myocardial perfusion due to CAD, or cardiomyopathy. Other causes include:

- ◆ diastolic dysfunction with preserved ejection fraction, impairment of ventricular filling by diminished relaxation or reduced compliance seen with hypertrophic cardiomyopathy, myocardial hypertrophy, and pericardial restriction
- ◆ mechanical disturbances in ventricular filling during diastole when there's too little blood for the ventricle to pump, as in mitral stenosis

secondary to rheumatic heart disease or constrictive pericarditis and atrial fibrillation

- ◆ systolic hemodynamic disturbances, such as excessive cardiac workload due to volume overload or pressure overload that limit the heart's pumping ability. These disturbances can result from mitral or aortic insufficiency, which causes volume overloading, and aortic stenosis or systemic hypertension, which result in increased resistance to ventricular emptying.

Heart failure affects 5.7 million people in the United States. It becomes more common with advancing age. Although heart failure may be acute (as a direct result of MI), it's generally a chronic disorder associated with sodium and water retention by the kidneys. Advances in diagnostic and therapeutic techniques have greatly improved the outlook for patients with heart failure, but the prognosis still depends on the underlying cause and its response to treatment. Fifty percent will die within 5 years of diagnosis.

## **Pathophysiology**

Reduced cardiac output triggers compensatory mechanisms, such as ventricular dilation, hypertrophy, increased sympathetic activity, and activation of the renin–angiotensin–aldosterone system. These mechanisms improve cardiac output at the expense of increased ventricular work. In cardiac dilation, an increase in end-diastolic ventricular volume (preload) causes increased stroke work and stroke volume during contraction, stretching cardiac muscle fibers beyond optimum limits and producing pulmonary congestion and pulmonary hypertension, which in turn lead to right-sided heart failure.

In ventricular hypertrophy, an increase in muscle mass or diameter of the left ventricle allows the heart to pump against increased resistance (impedance) to the outflow of blood. An increase in ventricular diastolic pressure necessary to fill the enlarged ventricle may compromise diastolic coronary blood flow, limiting oxygen supply to the ventricle and causing ischemia and impaired muscle contractility.

Increased sympathetic activity occurs as a response to decreased cardiac output and blood pressure by enhancing PVR, contractility, heart rate, and venous return. Signs of increased sympathetic activity, such as cool extremities and clamminess, may indicate impending heart failure. Increased sympathetic activity also restricts blood flow to the kidneys,

which respond by reducing the glomerular filtration rate and increasing tubular reabsorption of salt and water, in turn expanding the circulating blood volume. This renal mechanism, if unchecked, can aggravate congestion and produce overt edema.

Chronic heart failure may worsen as a result of respiratory tract infections, pulmonary embolism, stress, increased sodium or water intake, or failure to adhere to the prescribed treatment regimen.

## Complications

- ◆ Pulmonary edema
- ◆ Multiorgan failure
- ◆ Myocardial infarction

## Signs and Symptoms

Left-sided heart failure primarily produces pulmonary signs and symptoms; right-sided heart failure, primarily systemic signs and symptoms. However, heart failure often affects both sides of the heart.

Clinical signs of left-sided heart failure include dyspnea, orthopnea, crackles, possibly wheezing, hypoxia, respiratory acidosis, cough, cyanosis or pallor, palpitations, arrhythmias, elevated blood pressure, and pulsus alternans.

Clinical signs of right-sided heart failure include dependent peripheral edema, hepatomegaly, splenomegaly, JVD, ascites, slow weight gain, arrhythmias, positive hepatojugular reflex, abdominal distention, nausea, vomiting, anorexia, weakness, fatigue, dizziness, and syncope.



**ALERT** Excessive fluid can accumulate in the pericardium, requiring removal through pericardiocentesis.

## Diagnosis

- ◆ ECG may reflect heart strain or enlargement, ischemia, or old MI. It may also reveal atrial enlargement, tachycardia, and extrasystoles.
- ◆ Chest X-ray shows increased pulmonary vascular markings, interstitial edema, or pleural effusion and cardiomegaly.
- ◆ PAP monitoring typically demonstrates elevated pulmonary artery and PAWPs elevated; left ventricular end-diastolic pressure in left-sided heart

failure; and elevated right atrial pressure or CVP in right-sided heart failure.

- ◆ B-type natriuretic peptide (BNP) is a neurohormone produced predominantly by the ventricles and released in response to blood volume expansion or pressure overload. Blood concentrations greater than 100 pg/mL can be an accurate predictor of acute heart failure.
- ◆ Echocardiogram may demonstrate wall motion abnormalities and chamber dilation.

Other tests that may also demonstrate enlargement of the heart or decreased functioning include chest computed tomography scan, cardiac MRI, or nuclear scans, such as MUGA and radionuclide ventriculography.

## Treatment

The goal of therapy is to improve pump function by reversing the compensatory mechanisms producing the clinical effects, underlying disorders, and precipitating factors. Heart failure can be quickly controlled by treatment consisting of:

- ◆ ACE inhibitors to decrease PVR
- ◆ antiembolism stockings to reduce the risk of venostasis and thromboembolus formation
- ◆ bed rest for acute heart failure
- ◆ carvedilol, a nonselective BB with alpha-receptor blockade to reduce mortality and improve quality of life
- ◆ digoxin or dopamine to strengthen myocardial contractility
- ◆ diuresis to reduce total blood volume and circulatory congestion
- ◆ inotropic agents, such as dobutamine and milrinone, given I.V. to improve the heart's ability to pump
- ◆ nesiritide, a recombinant form of endogenous human BNP, to reduce sodium through its diuretic action
- ◆ vasodilators to increase cardiac output by reducing the impedance to ventricular outflow (afterload).

Excess fluid can be removed through dialysis if necessary. Circulatory assistance can be provided by implanted devices, such as the IABP and the left ventricular assist device (LVAD), but they're only temporary solutions.

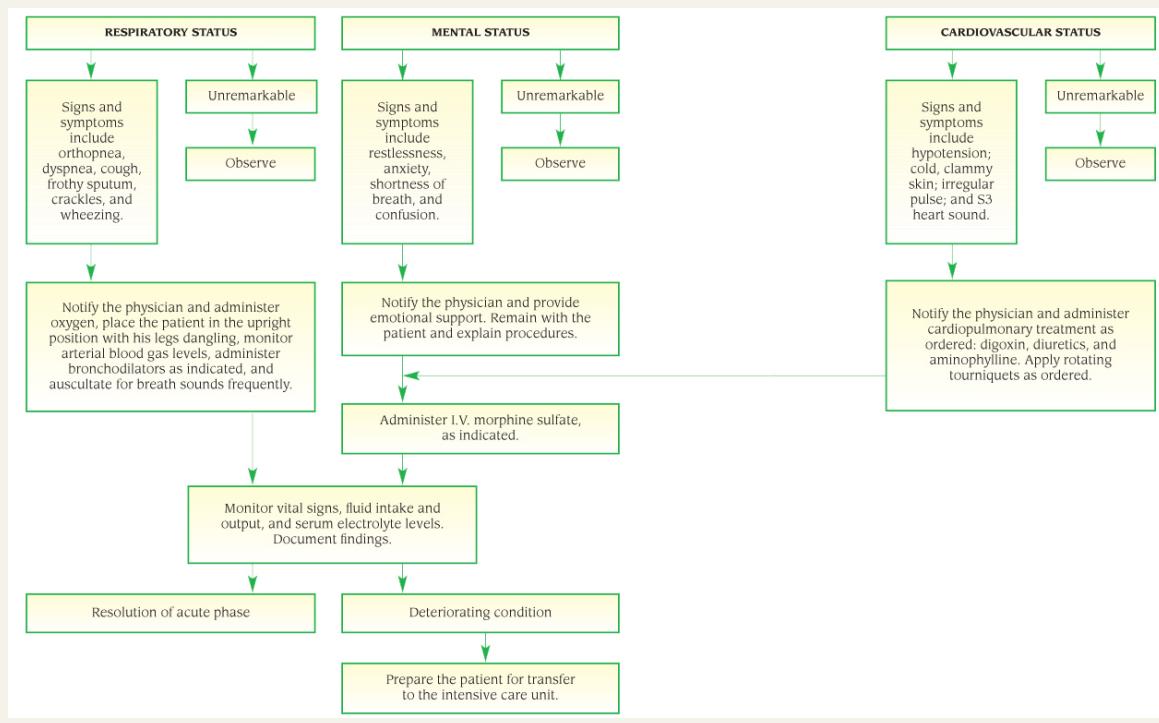
A LVAD may be an option for those with refractory heart failure who are not eligible for transplant. This procedure may increase the patient's quality

of life.

Watch for and treat complications, which typically may include pulmonary edema (See *Pulmonary edema: How to intervene*, pages 48 and 49); venostasis, with predisposition to thromboembolism (associated primarily with prolonged bed rest); cerebral insufficiency; and renal insufficiency, with severe electrolyte imbalance.

## Pulmonary Edema: How To Intervene

Obtain the patient history; assist with diagnostic tests; and assess respiratory, mental, and cardiovascular status.



## Special Considerations

During the acute phase of heart failure:

- ◆ Give supplemental oxygen to help make breathing easier.
- ◆ Weigh the patient daily and check for peripheral edema. Carefully monitor I.V. intake and urine output, vital signs, and mental status.

Auscultate the heart for abnormal sounds ( $S_3$  gallop) and the lungs for crackles or rhonchi. Report changes at once.

- ◆ Give the patient a fluid restriction of less than 2 L/day from all sources.
- ◆ Frequently monitor BUN, creatinine, and serum potassium, sodium, chloride, and magnesium levels.
- ◆ Make sure the patient has continuous cardiac monitoring during acute and advanced stages to identify and treat arrhythmias promptly.
- ◆ To reduce the risk of deep vein thrombosis (DVT) due to vascular congestion, assist the patient with range-of-motion exercises. Enforce bed rest and apply antiembolism stockings.
- ◆ Allow adequate rest periods.

To prepare the patient for discharge:

- ◆ Advise the patient to avoid foods high in sodium, such as canned or commercially prepared foods and dairy products, to curb fluid overload.
- ◆ Encourage participation in an outpatient cardiac rehabilitation program.
- ◆ Explain to the patient that the potassium lost through diuretic therapy may need to be replaced by taking a prescribed potassium supplement and eating high-potassium foods, such as bananas and apricots.
- ◆ Stress the need for regular checkups.
- ◆ Stress the importance of taking digoxin exactly as prescribed. Tell the patient to watch for and immediately report signs of toxicity, such as anorexia, vomiting, and yellow vision.
- ◆ Tell the patient to notify the practitioner promptly if his or her pulse is unusually irregular or measures less than 60 beats/minute; if experiencing dizziness, blurred vision, shortness of breath, a persistent dry cough, palpitations, increased fatigue, paroxysmal nocturnal dyspnea, swollen ankles, or decreased urine output; or if he notices rapid weight gain (3 to 5 lb [1.4 to 2.3 kg] in 1 week).



## PREVENTION

- ◆ *Instruct patient to make lifestyle modifications, including regular exercise, weight loss, smoking cessation, stress reduction, and reduced sodium, alcohol, and fat intake.*
- ◆ *Instruct patient to practice compliance with and timely administration of maintenance doses of diuretics and cardiac drugs.*

## DILATED CARDIOMYOPATHY

### Causes and Incidence

Dilated cardiomyopathy results from extensively damaged myocardial muscle fibers. It is the most common type of cardiomyopathy. This disorder interferes with myocardial metabolism and grossly dilates all four chambers of the heart, giving the heart a globular appearance and shape. In this disorder, hypertrophy may be present. Dilated cardiomyopathy leads to intractable heart failure, arrhythmias, and emboli. Because this disease isn't usually diagnosed until it's in the advanced stages, the patient's prognosis is generally poor.

The cause of most cardiomyopathies is unknown. Occasionally, dilated cardiomyopathy results from myocardial destruction by toxic, infectious, or metabolic agents, such as certain viruses, endocrine and electrolyte disorders, nutritional deficiencies, and certain cardiotoxic anticancer drugs (e.g., doxorubicin). Other causes include muscle disorders (myasthenia gravis, progressive muscular dystrophy, and myotonic dystrophy), infiltrative disorders (hemochromatosis and amyloidosis), and sarcoidosis.

Cardiomyopathy may also be a complication of alcoholism. In such cases, it may improve with abstinence from alcohol but recurs when the patient resumes drinking.

Dilated cardiomyopathy occurs in 1 in 2,500 people and affects all ages and both sexes. It's most common in adult men between the ages of 30 and 40.

### Pathophysiology

How viruses induce cardiomyopathy is unclear, but researchers suspect a link between viral myocarditis and subsequent dilated cardiomyopathy, especially after infection with poliovirus, coxsackievirus B, influenza virus, or human immunodeficiency virus.

Metabolic cardiomyopathies are related to endocrine and electrolyte disorders and nutritional deficiencies. Thus, dilated cardiomyopathy may develop in patients with hyperthyroidism, pheochromocytoma, beriberi (thiamine deficiency), or kwashiorkor (protein deficiency). Cardiomyopathy may also result from rheumatic fever, especially among children with myocarditis.

Antepartal or postpartal cardiomyopathy may develop during the last trimester or within months after delivery. Its cause is unknown, but it occurs most frequently in multiparous women older than 30, particularly those with malnutrition or preeclampsia. In these patients, cardiomegaly and heart failure may reverse with treatment, allowing a subsequent normal pregnancy. If cardiomegaly persists despite treatment, the prognosis is poor.

## Complications

- ◆ Heart failure
- ◆ Arrhythmias
- ◆ Emboli
- ◆ Ventricular arrhythmias
- ◆ Syncope
- ◆ Sudden death

## Signs and Symptoms

In dilated cardiomyopathy, the heart ejects blood less efficiently than normal. Consequently, a large volume of blood remains in the left ventricle after systole, causing signs of heart failure—both left-sided (shortness of breath, orthopnea, dyspnea on exertion, paroxysmal nocturnal dyspnea, fatigue, and an irritating dry cough at night) and right-sided (edema, liver engorgement, and JVD). Dilated cardiomyopathy also produces peripheral cyanosis and sinus tachycardia or atrial fibrillation at rest in some patients secondary to low cardiac output. Auscultation reveals diffuse apical impulses, pansystolic murmur (mitral and tricuspid insufficiency secondary to cardiomegaly and weak papillary muscles), and S<sub>3</sub> and S<sub>4</sub> gallop rhythms.

## Diagnosis

Diagnosis of dilated cardiomyopathy requires elimination of other possible causes of heart failure and arrhythmias.

- ◆ ECG and angiography rule out ischemic heart disease; ECG may also show biventricular hypertrophy, sinus tachycardia, atrial enlargement, and, in 20% of patients, atrial fibrillation and bundle branch block.
- ◆ Chest X-ray shows cardiomegaly—usually affecting all heart chambers—and may demonstrate pulmonary congestion, pleural or pericardial effusion, or pulmonary venous hypertension.

- ◆ Chest computed tomography scan or echocardiography identifies left ventricular thrombi, global hypokinesia, and degree of left ventricular dilation.
- ◆ Nuclear heart scans, such as MUGA and ventriculography, show heart enlargement, lung congestion, heart failure, and decreased movement or functioning of the heart.

## **Treatment**

Therapeutic goals include correcting the underlying causes and improving the heart's pumping ability with digoxin, diuretics, oxygen, and a sodium-restricted diet. Other options include an ACE inhibitor or ARB, a beta-blocker, aspirin, and, potentially, other blood-thinning medications. Vasodilators reduce preload and afterload, thereby decreasing congestion and increasing cardiac output. Acute heart failure requires vasodilation with nitroprusside or nitroglycerin I.V.

When these treatments fail, therapy may require pacemakers, implantable cardiac defibrillations, LVADs, or, as a last resort, a heart transplant for carefully selected patients.

## **Special Considerations**

In the patient with acute failure:

- ◆ Monitor for signs of progressive failure (increasing crackles and dyspnea and increased JVD) and compromised renal perfusion (oliguria, elevated BUN and creatinine levels, and electrolyte imbalances). Weigh the patient daily.
- ◆ If the patient is receiving vasodilators, check blood pressure and heart rate. If the patient becomes hypotensive, stop the infusion and place in a supine position, with legs elevated to increase venous return and to ensure cerebral blood flow.
- ◆ If the patient is receiving diuretics, monitor for signs of resolving congestion (decreased crackles and dyspnea) or too-vigorous diuresis. Check serum potassium level for hypokalemia, especially if therapy includes digoxin.
- ◆ Therapeutic restrictions and an uncertain prognosis usually cause profound anxiety and depression, so offer support and let the patient express his or her feelings. Be flexible with visiting hours.

- ◆ Before discharge, teach the patient about their illness and its treatment. Emphasize the need to avoid alcohol and smoking, to restrict sodium intake, to watch for weight gain (a weight gain of 3 lb [1.4 kg] over 1 to 2 days indicates fluid accumulation), and to take digoxin as prescribed, watching for its adverse effects (anorexia, nausea, vomiting, and yellow vision).
- ◆ Encourage family members to learn cardiopulmonary resuscitation (CPR).

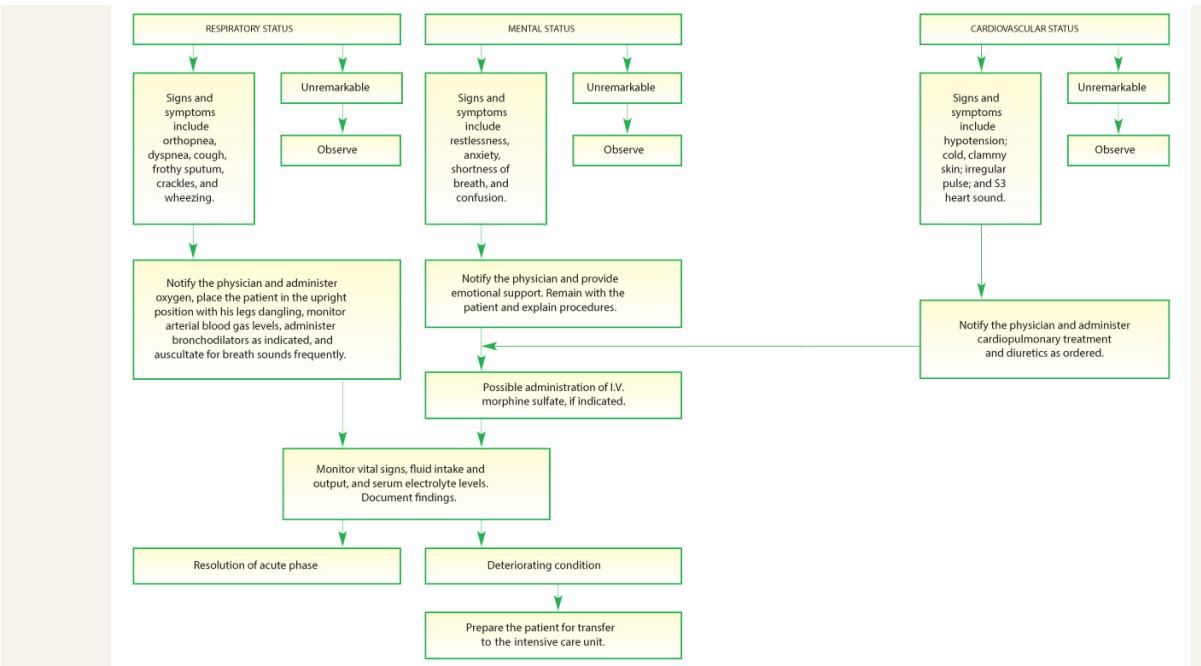
## HYPERTROPHIC CARDIOMYOPATHY

### Causes and Incidence

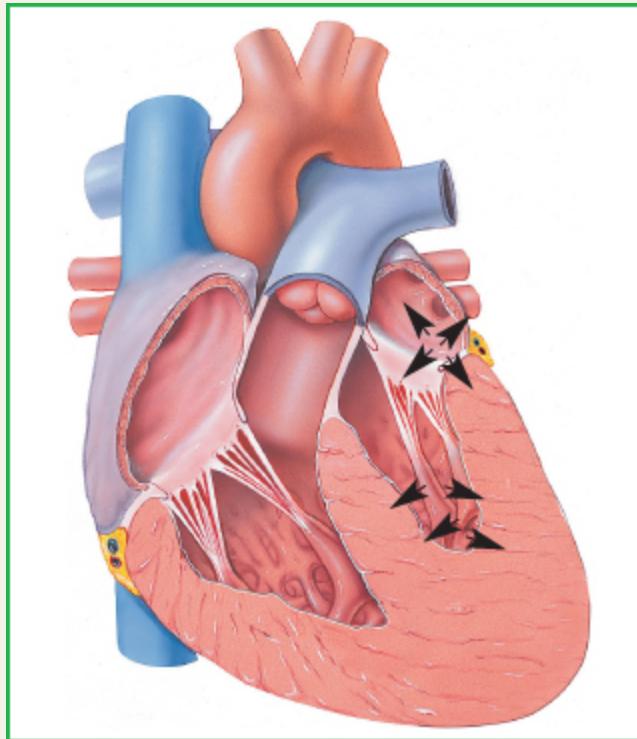
This primary disease of cardiac muscle, also called *idiopathic hypertrophic subaortic stenosis*, is characterized by disproportionate, asymmetrical thickening of the interventricular septum, particularly in the left ventricle's free wall. In hypertrophic cardiomyopathy, cardiac output may be low, normal, or high, depending on whether the stenosis is obstructive or nonobstructive. If cardiac output is normal or high, the disorder may go undetected for years; but low cardiac output may lead to potentially fatal heart failure. The disease course varies; some patients progressively deteriorate; others remain stable for years. (See *Looking at hypertrophic cardiomyopathy*, pages 50 to 52.)

### Looking At Hypertrophic Cardiomyopathy

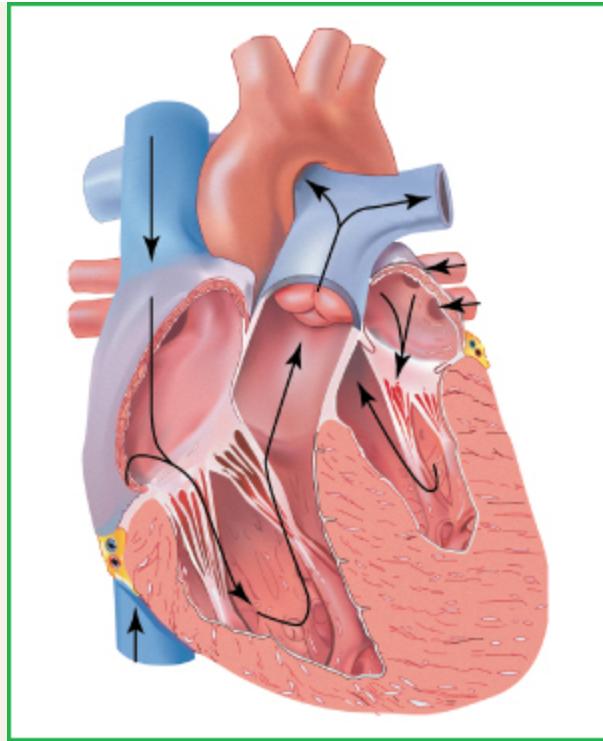
1. The left ventricle and interventricular septum hypertrophy and become stiff, noncompliant, and unable to relax during ventricular filling.



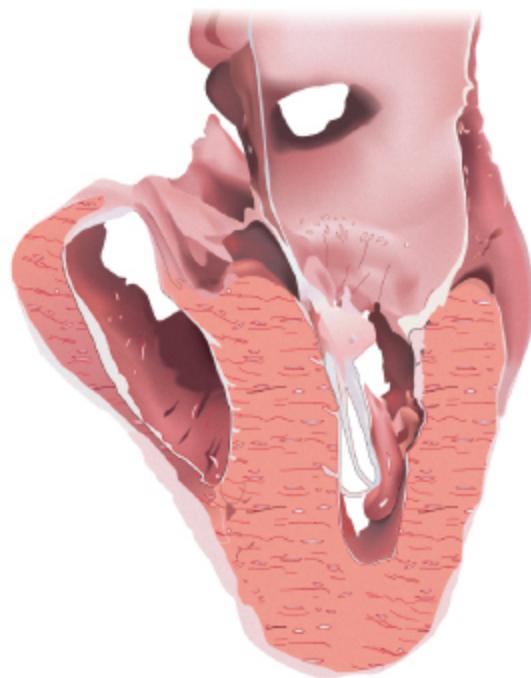
- As the ventricle's ability to fill decreases, the pressure increases, and left atrial and pulmonary venous pressures rise.



- The left ventricle forcefully contracts but can't sufficiently relax.



4. The anterior leaflet of the mitral valve is drawn toward the interventricular septum as the blood is forcefully ejected. Early closure of the outflow tract results because of the decreasing ejection fraction.



This disorder affects 1 in 500 people and is more common in men than in women and more so in blacks than in whites. It is also usually the cause of sudden death, particularly in otherwise healthy athletes.

### **Pathophysiology**

Despite being designated as idiopathic, hypertrophic cardiomyopathy may be inherited as a non-sex-linked autosomal dominant trait in almost all cases. Most patients have obstructive disease, resulting from effects of ventricular septal hypertrophy and the movement of the anterior mitral valve leaflet into the outflow tract during systole. Eventually, left ventricular dysfunction, from rigidity and decreased compliance, causes pump failure.

### **Complications**

- ◆ Pulmonary hypertension
- ◆ Heart failure
- ◆ Ventricular arrhythmias
- ◆ Sudden death

## **Signs and Symptoms**

Clinical features of the disorder may not appear until it's well advanced, when atrial dilation and, possibly, atrial fibrillation abruptly reduce blood flow to the left ventricle. Reduced inflow and subsequent low output may produce angina pectoris, arrhythmias, dyspnea, orthopnea, syncope, heart failure, and death. Auscultation reveals a medium-pitched systolic ejection murmur along the left sternal border and at the apex; palpation reveals a peripheral pulse with a characteristic double impulse (pulsus bisferiens) and, with atrial fibrillation, an irregular pulse.

## **Diagnosis**

Diagnosis depends on typical clinical findings and these test results:

- ◆ Echocardiography (most useful) shows increased thickness of the intraventricular septum and abnormal motion of the anterior mitral leaflet during systole, occluding left ventricular outflow in obstructive disease.
- ◆ Cardiac catheterization reveals elevated left ventricular end-diastolic pressure and, possibly, mitral insufficiency.
- ◆ ECG usually shows left ventricular hypertrophy, T-wave inversion, left anterior hemiblock, Q waves in precordial and inferior leads, ventricular arrhythmias, and, possibly, atrial fibrillation.
- ◆ Auscultation confirms an early systolic murmur.

## **Treatment**

The goals of treatment are to relax the ventricle and to relieve outflow tract obstruction. Agents such as metoprolol, a BB, slow heart rate and increase ventricular filling by relaxing the obstructing muscle, thereby reducing angina, syncope, dyspnea, and arrhythmias. Atrial fibrillation necessitates cardioversion to treat the arrhythmia and, because of the high risk of systemic embolism, anticoagulant therapy until fibrillation subsides. Because vasodilators such as nitroglycerin reduce venous return by permitting pooling of blood in the periphery, decreasing ventricular volume and chamber size, and may cause further obstruction, they're contraindicated in patients with hypertrophic cardiomyopathy. Also contraindicated are ACE inhibitors and ARBs because they can also worsen the obstruction. Disopyramide is preferred because of its negative inotropic

properties. Patients with potentially lethal arrhythmias may need an implantable cardioverter–defibrillator to prevent sudden death.

If drug therapy fails, surgery is indicated. Septal myectomy reduces the thickness of the septum through removal of a portion of the proximal septum. This can be performed alone or in combination with mitral valve replacement, which may ease outflow tract obstruction and relieve symptoms. However, complications, such as complete heart block and VSD, can occur, requiring pacemaker implantation.

## Special Considerations

- ◆ Because syncope or sudden death may follow well-tolerated exercise, warn such patients against strenuous physical activity such as running.
- ◆ Administer medications as prescribed. *Caution:* Avoid nitroglycerin, digoxin, and diuretics because they can worsen obstruction. Warn the patient not to stop taking their beta-blocker abruptly, because doing so may increase myocardial demands. To determine the patient's tolerance for an increased dosage of the beta-blocker, take his or her pulse to check for bradycardia. Also take a blood pressure reading while the patient is supine and while standing (a drop in blood pressure [ $>10$  mm Hg] when standing may indicate orthostatic hypotension).
- ◆ Before dental work or surgery, tell the patient to discuss prophylaxis for subacute infective endocarditis with his or her healthcare provider.
- ◆ Provide psychological support. If the patient is hospitalized for a long time, be flexible with visiting hours and encourage occasional weekends away from the hospital, if possible. Refer the patient for psychosocial counseling to help the patient and family accept restricted lifestyle and poor prognosis.
- ◆ If the patient is a child, have the parents arrange for them to continue school work or studies in the healthcare facility.
- ◆ Because sudden cardiac arrest is possible, urge the patient's family to learn cardiopulmonary resuscitation.

## Cardiac Complications

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### HYPOVOLEMIC SHOCK

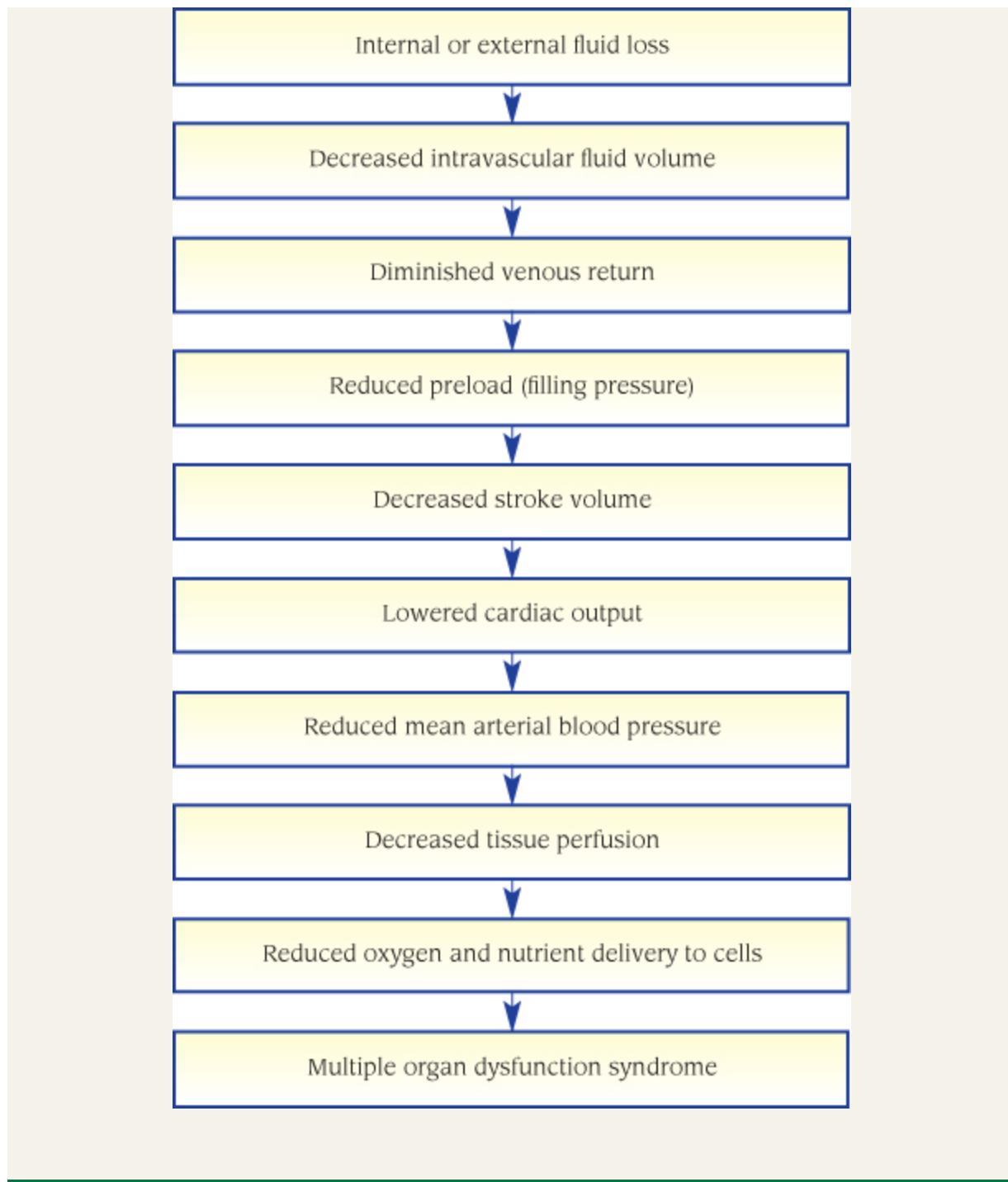
## Causes and Incidence

In hypovolemic shock, reduced intravascular blood volume causes circulatory dysfunction and inadequate tissue perfusion. Without sufficient blood or fluid replacement, hypovolemic shock syndrome may lead to irreversible cerebral and renal damage, cardiac arrest, and, ultimately, death. Hypovolemic shock requires early recognition of signs and symptoms and prompt, aggressive treatment to improve the prognosis. (See *What happens in hypovolemic shock*, page 55.)



## PATHOPHYSIOLOGY WHAT HAPPENS IN HYPOVOLEMIC SHOCK

In hypovolemic shock, vascular fluid volume loss causes extreme tissue hypoperfusion. Internal fluid losses can result from hemorrhage or third-space fluid shifting. External fluid loss can result from severe bleeding or from severe diarrhea, diuresis, or vomiting. Inadequate vascular volume leads to decreased venous return and cardiac output. The resulting drop in arterial blood pressure activates the body's compensatory mechanisms in an attempt to increase vascular volume. If compensation is unsuccessful, decompensation and death may occur.



Hypovolemic shock usually results from acute blood loss—about one fifth of total volume. Such massive blood loss may result from GI bleeding, internal hemorrhage (hemothorax and hemoperitoneum), external hemorrhage (accidental or surgical trauma), or from any condition that reduces circulating intravascular plasma volume or other body fluids such

as in severe burns. Other underlying causes of hypovolemic shock include intestinal obstruction, peritonitis, acute pancreatitis, ascites and dehydration from excessive perspiration, severe diarrhea or protracted vomiting, diabetes insipidus, diuresis, or inadequate fluid intake.

## **Pathophysiology**

Hypovolemic shock occurs when there is not enough intravascular volume to maintain tissue perfusion. At first, the body responds by stimulating thirst and reducing the amount of fluid filtered through the kidneys. Additionally, heart rate becomes elevated to increase cardiac output, contractility increases to maintain stroke volume, and increased systemic vascular resistance shunts blood away from the periphery and toward the heart and central nervous system. Hypotension is a late sign since blood pressure can be maintained initially through compensatory vasoconstriction, which decreases tissue perfusion. Cardiac arrest may occur soon after.

## **Complications**

- ◆ Acute respiratory distress syndrome
- ◆ Acute tubular necrosis
- ◆ Disseminated intravascular coagulation (DIC)
- ◆ Multiple-organ-dysfunction syndrome

## **Signs and Symptoms**

Hypovolemic shock produces a syndrome of hypotension, with narrowing pulse pressure; decreased sensorium; tachycardia; rapid, shallow respirations; reduced urine output (<25 mL/hour); and cold, pale, clammy skin. Metabolic acidosis with an accumulation of lactic acid develops as a result of tissue anoxia, as cellular metabolism shifts from aerobic to anaerobic pathways. DIC is a possible complication of hypovolemic shock.

## **Diagnosis**

No single symptom or diagnostic test establishes the diagnosis or severity of shock. Characteristic laboratory findings include:

- ◆ elevated potassium, serum lactate, and BUN levels
- ◆ increased urine specific gravity ( $>1.020$ ) and urine osmolality

- ◆ decreased blood pH and partial pressure of arterial oxygen and increased partial pressure of arterial carbon dioxide.

In addition, gastroscopy, aspiration of gastric contents through a nasogastric (NG) tube, computed tomography scan, and X-rays identify internal bleeding sites; coagulation studies may detect coagulopathy from DIC. Echocardiography or right heart catheterization can help differentiate between hypovolemic and cardiogenic shock.

## Treatment

Emergency treatment measures must include prompt and adequate blood and fluid replacement to restore intravascular volume and raise blood pressure. Saline solution or lactated Ringer solution, then possibly plasma proteins (albumin) or other plasma expanders, may produce adequate volume expansion until whole blood can be matched. A rapid solution infusion system of 1 to 2 L of isotonic crystalloids can be used and continued if hypotension persists. Measuring CVP can assist in directing therapy. Treatment may also include oxygen administration, identification of bleeding site, control of bleeding by direct measures (such as application of pressure and elevation of a limb) and, possibly, surgery.

## Special Considerations

Management of hypovolemic shock necessitates prompt, aggressive supportive measures and careful assessment and monitoring of vital signs. Follow these priorities:

- ◆ Check for a patent airway and adequate circulation. If blood pressure and heart rate are absent, start CPR.
- ◆ Record blood pressure, pulse rate, peripheral pulses, respiratory rate, and other vital signs every 15 minutes and the electrocardiograph continuously. Systolic blood pressure lower than 80 mm Hg usually results in inadequate coronary artery blood flow, cardiac ischemia, arrhythmias, and further complications of low cardiac output. When blood pressure drops below 80 mm Hg, increase the oxygen flow rate and notify the practitioner immediately. A progressive drop in blood pressure, accompanied by a thready pulse, generally signals inadequate cardiac output from reduced intravascular volume. Notify the practitioner and increase the infusion rate.

- ◆ Start I.V. lines with normal saline or lactated Ringer solution, using a large-bore catheter (14G), which allows easier administration of later blood transfusions. (*Caution:* Don't start I.V. lines in the legs of a patient in shock who has suffered abdominal trauma, because infused fluid may escape through the ruptured vessel into the abdomen.)
- ◆ An indwelling urinary catheter may be inserted to measure hourly urine output. If output is less than 30 mL/hour in adults, increase the fluid infusion rate, but watch for signs of fluid overload such as an increase in PAWP. Notify the practitioner if urine output doesn't improve. An osmotic diuretic such as mannitol may be ordered to increase renal blood flow and urine output. Determine how much fluid to give by checking blood pressure, urine output, CVP, or PAWP. (To increase accuracy, CVP should be measured at the level of the right atrium, using the same reference point on the chest each time.)
- ◆ Draw an arterial blood sample to measure blood gas levels. Administer oxygen by face mask or airway to ensure adequate oxygenation of tissues. Adjust the oxygen flow rate to a higher or lower level, as blood gas measurements indicate.
- ◆ Draw venous blood for complete blood count and electrolyte, type and crossmatch, and coagulation studies.
- ◆ During therapy, assess skin color and temperature, and note changes. Cold, clammy skin may be a sign of continuing peripheral vascular constriction, indicating progressive shock.
- ◆ Watch for signs of impending coagulopathy (petechiae, bruising, and bleeding or oozing from gums or venipuncture sites).
- ◆ Explain procedures and their purpose. Throughout these emergency measures, provide emotional support to the patient and family.



### PREVENTION

- *Recognize patients with conditions that reduce blood volume as at-risk patients.*
- *Estimate fluid loss and replace, as necessary, to prevent hypovolemic shock.*

## CARDIOGENIC SHOCK

## Causes and Incidence

Sometimes called *pump failure*, cardiogenic shock is a condition of diminished cardiac output that severely impairs tissue perfusion. It reflects severe left-sided heart failure and occurs as a serious complication in 5% to 10% of all patients hospitalized with acute MI. Cardiogenic shock can result from any condition that causes significant left ventricular dysfunction with reduced cardiac output, such as MI (most common), myocardial ischemia, papillary muscle dysfunction, or end-stage cardiomyopathy. The incidence of cardiogenic shock is higher those with an elevated body mass index, and also higher in men than in women because of their higher incidence of CAD. Historically, mortality for cardiogenic shock had been 80% to 90%, but recent studies indicate that the rate has dropped to 40% to 50% due to improved interventional procedures and better therapies. Mortality is expected to decline even further.

## Pathophysiology

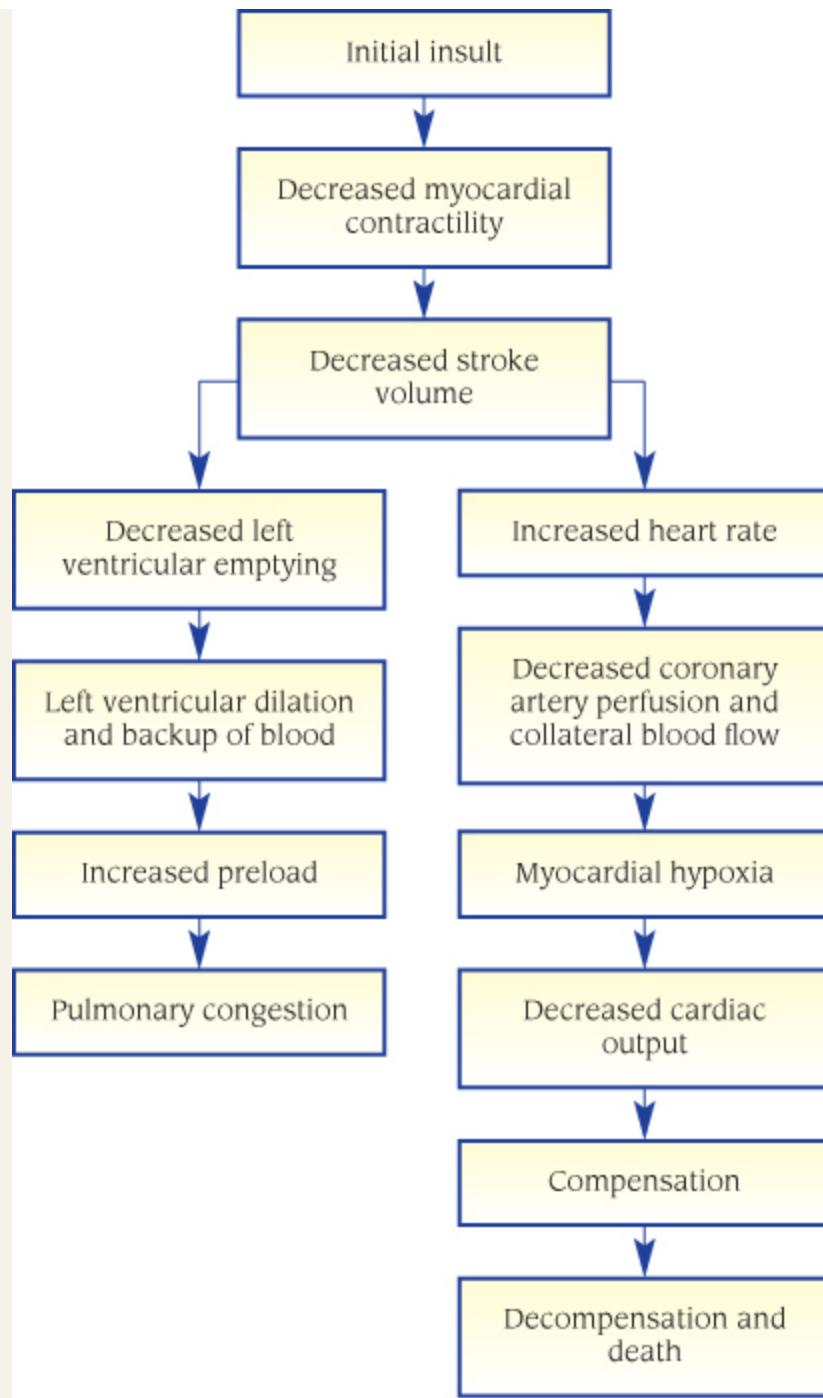
Regardless of the underlying cause, left ventricular dysfunction sets into motion a series of compensatory mechanisms that attempt to increase cardiac output and, in turn, maintain vital organ function. (See *What happens in cardiogenic shock*, page 57.) As cardiac output falls in left ventricular dysfunction, aortic and carotid baroreceptors initiate sympathetic nervous responses, which increase heart rate, left ventricular filling pressure, and peripheral resistance to flow, to enhance venous return to the heart. These compensatory responses initially stabilize the patient but later cause deterioration with the rising oxygen demands of the already compromised myocardium. These events comprise a vicious circle of low cardiac output, sympathetic compensation, myocardial ischemia, and even lower cardiac output.



### PATHOPHYSIOLOGY WHAT HAPPENS IN CARDIOGENIC SHOCK

When the myocardium can't contract sufficiently to maintain adequate cardiac output, stroke volume decreases, and the heart can't eject an adequate volume of blood with each contraction. The blood backs up behind the weakened left ventricle, increasing preload and

causing pulmonary congestion. In addition, to compensate for the drop in stroke volume, the heart rate increases in an attempt to maintain cardiac output. As a result of the diminished stroke volume, coronary artery perfusion and collateral blood flow decrease. All of these mechanisms increase the heart's workload and enhance left-sided heart failure. The result is myocardial hypoxia, further decreased cardiac output, and a triggering of compensatory mechanisms to prevent decompensation and death.



## Signs and Symptoms

Cardiogenic shock produces signs of poor tissue perfusion: cold, pale, clammy skin; a decrease in systolic blood pressure to 30 mm Hg below baseline, or a sustained reading below 80 mm Hg not attributable to

medication; tachycardia; rapid, shallow respirations; oliguria (<20 mL/hour); restlessness; mental confusion and obtundation; narrowing pulse pressure; and cyanosis. Although many of these clinical features also occur in heart failure and other shock syndromes, they're usually more profound in cardiogenic shock.

## Diagnosis

Auscultation may detect gallop rhythm, faint heart sounds, and, possibly, if the shock results from rupture of the ventricular septum or papillary muscles, a holosystolic murmur.

- ◆ PAP monitoring may show increased PAP and increased PAWP, reflecting a rise in left ventricular end-diastolic pressure (preload) and increased resistance to left ventricular emptying (afterload) due to ineffective pumping and increased PVR. Thermodilution technique measures decreased cardiac output.
- ◆ Invasive arterial pressure monitoring may indicate hypotension due to impaired ventricular ejection.
- ◆ ABG analysis may show metabolic acidosis and hypoxia.
- ◆ ECG may show possible evidence of acute MI, ischemia, or ventricular aneurysm.
- ◆ Echocardiography can determine left ventricular function and reveal valvular abnormalities.
- ◆ Enzyme levels may show elevated CK, lactate dehydrogenase, aspartate aminotransferase, and alanine aminotransferase, which point to MI or ischemia and suggest heart failure or shock. Troponin I, troponin T, and CK may confirm acute MI.

Additional tests determine other conditions that can lead to pump dysfunction and failure, such as cardiac arrhythmias, cardiac tamponade, papillary muscle infarct or rupture, ventricular septal rupture, pulmonary emboli, venous pooling (associated with vasodilators and continuous intermittent positive-pressure breathing), and hypovolemia.

## Treatment

The aim of treatment is to enhance cardiovascular status by increasing cardiac output, improving myocardial perfusion, and decreasing cardiac workload with combinations of various cardiovascular drugs and

mechanical-assist techniques. Myocardial reperfusion can be accomplished by PTCA, stents, thrombolytic therapy, or bypass grafting. Drug therapy may include dopamine I.V., a vasopressor that increases cardiac output, blood pressure, and renal blood flow; milrinone or dobutamine I.V., inotropic agents that increase myocardial contractility; norepinephrine, when a more potent vasoconstrictor is necessary; and nitroprusside I.V., a vasodilator that may be used with a vasopressor to further improve cardiac output by decreasing PVR (afterload) and reducing left ventricular end-diastolic pressure (preload). However, the patient's blood pressure must be adequate to support nitroprusside therapy and must be monitored closely.

The IABP is a mechanical-assist device that attempts to improve coronary artery perfusion and decrease cardiac workload. (See *Understanding the IABP*, page 58.) The inflatable balloon pump is percutaneously or surgically inserted through the femoral artery into the descending thoracic aorta. The balloon inflates during diastole to increase coronary artery perfusion pressure and deflates before systole (before the aortic valve opens) to reduce resistance to ejection (afterload) and reduce cardiac workload. Improved ventricular ejection, which significantly improves cardiac output, and a subsequent vasodilation in the peripheral vasculature lead to lower preload volume.

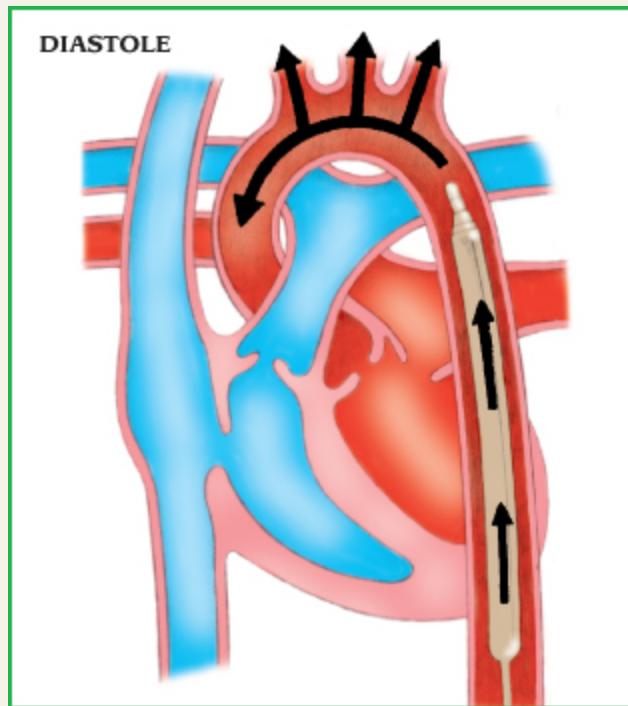
## **Understanding The IABP**

An intra-aortic balloon pump (IABP) consists of a polyurethane balloon attached to an external pump console by means of a large-lumen catheter. It is inserted percutaneously through the femoral artery and positioned in the descending aorta, just distal to the left subclavian artery and above the renal arteries.

### **Push...**

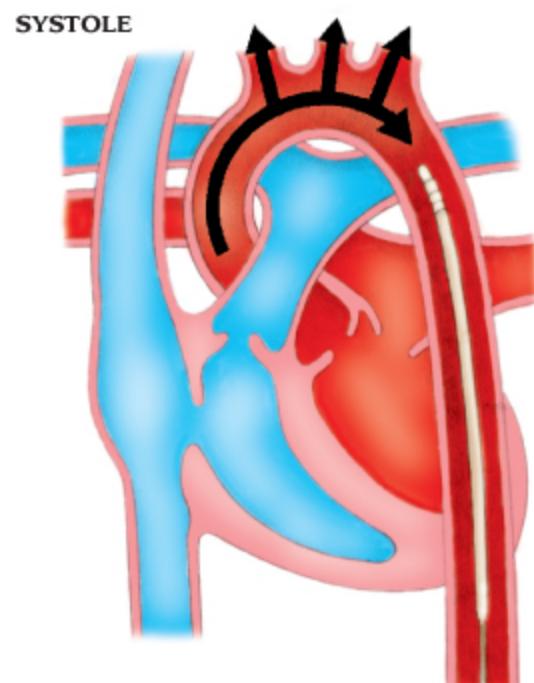
This external pump works in precise counterpoint to the left ventricle, inflating the balloon with helium early in diastole and deflating it just before systole. As the balloon inflates, it forces blood toward the aortic valve, thereby raising pressure in the aortic root and augmenting diastolic pressure to improve coronary perfusion. It also improves

peripheral circulation by forcing blood through the brachiocephalic, common carotid, and subclavian arteries arising from the aortic trunk.



### **...and Pull**

The balloon deflates rapidly at the end of diastole, creating a vacuum in the aorta. This reduces aortic volume and pressure, thereby decreasing the resistance to left ventricular ejection (afterload). This decreased workload, in turn, reduces the heart's oxygen requirements and, combined with the improved myocardial perfusion, helps prevent or diminish myocardial ischemia.



When drug therapy and IABP insertion fail, treatment may require the use of a ventricular assist device. This device (which may be either temporary or permanent) diverts systemic blood flow from a diseased ventricle into a centrifugal pump. It assists the heart's pumping action rather than replacing it.

### Special Considerations

- ◆ At the first sign of cardiogenic shock, check the patient's blood pressure and heart rate. If the patient is hypotensive or is having difficulty breathing, ensure a patent I.V. line and a patent airway, and provide oxygen to promote tissue oxygenation. Notify the practitioner immediately.
- ◆ Monitor ABG values to measure oxygenation and detect acidosis from poor tissue perfusion. Increase oxygen delivery as indicated. Check complete blood count and electrolyte levels.
- ◆ After diagnosis, monitor cardiac rhythm continuously and assess skin color, temperature, and other vital signs often. Watch for a drop in systolic blood pressure to less than 80 mm Hg (usually compromising cardiac output further). Report hypotension immediately.

- ◆ An indwelling urinary catheter may be inserted to measure urine output. Notify the practitioner if output drops below 30 mL/hour.
- ◆ Using a pulmonary artery catheter, closely monitor PAP, PAWP, and, if equipment is available, cardiac output. A high PAWP indicates heart failure and should be reported.
- ◆ When a patient is on the IABP, reposition the patient often and perform passive range-of-motion exercises to prevent skin breakdown. However, don't flex the patient's "ballooned" leg at the hip because this may displace or fracture the catheter. Assess pedal pulses and skin temperature and color to make sure circulation to the leg is adequate. Check the dressing on the insertion site frequently for bleeding, and change it according to facility protocol. Also, check the site for hematoma or signs of infection, and culture any drainage.
- ◆ After the patient becomes hemodynamically stable, the frequency of balloon inflation is gradually reduced to wean from the IABP. During weaning, carefully watch for monitor changes, chest pain, and other signs of recurring cardiac ischemia and shock.
- ◆ Provide psychological support and reassurance because the patient and family may be anxious about the ICU, IABP, and other tubes and devices. To ease emotional stress, plan your care to allow frequent rest periods, and provide as much privacy as possible.



**PREVENTION** Emphasize the fact that prevention requires timely, thorough, and aggressive identification and treatment of causative disorders.

## VENTRICULAR ANEURYSM

### Causes and Incidence

A ventricular aneurysm is an outpouching, almost always of the left ventricle, that produces ventricular wall dysfunction in about 25% of patients after MI. Ventricular aneurysm may develop within weeks after MI. Untreated ventricular aneurysm can lead to arrhythmias, systemic embolization, or heart failure and may cause sudden death. Resection improves the prognosis in patients with heart failure or refractory patients who have developed ventricular arrhythmias.

## **Pathophysiology**

When MI destroys a large muscular section of the left ventricle, necrosis reduces the ventricular wall to a thin sheath of fibrous tissue. Under intracardiac pressure, this thin layer stretches and forms a separate noncontractile sac (aneurysm). Abnormal muscular wall movement accompanies ventricular aneurysm and includes akinesia (lack of movement), dyskinesia (paradoxical movement), asynergia (decreased and inadequate movement), and asynchrony (uncoordinated movement). During systolic ejection, the abnormal muscular wall movements associated with the aneurysm cause the remaining normally functioning myocardial fibers to increase the force of contraction to maintain stroke volume and cardiac output. At the same time, a portion of the stroke volume is lost to passive distention of the noncontractile sac.

## **Complications**

- ◆ Ventricular arrhythmias
- ◆ Cerebral embolism
- ◆ Heart failure
- ◆ Death

## **Signs and Symptoms**

Ventricular aneurysm may cause arrhythmias—such as premature ventricular contractions or ventricular tachycardia—palpitations, signs of cardiac dysfunction (weakness on exertion, fatigue, and angina) and, occasionally, a visible or palpable systolic precordial bulge. This condition may also lead to left ventricular dysfunction, with chronic heart failure (dyspnea, fatigue, edema, crackles, gallop rhythm, and JVD); pulmonary edema; systemic embolization; and, with left-sided heart failure, pulsus alternans. Ventricular aneurysms enlarge but seldom rupture.

## **Diagnosis**

Persistent ventricular arrhythmias, onset of heart failure, or systemic embolization in a patient with left-sided heart failure and a history of MI strongly suggest ventricular aneurysm. Indicative tests include the following:

- ◆ Left ventriculography during catheterization reveals left ventricular enlargement, with an area of akinesia or dyskinesia and diminished cardiac function.
- ◆ ECG may show persistent ST–T wave elevations after infarction.
- ◆ Chest X-ray may demonstrate an abnormal bulge distorting the heart's contour if the aneurysm is large; the X-ray may be normal if the aneurysm is small.
- ◆ Noninvasive nuclear cardiology scan may indicate the site of infarction and suggest the area of aneurysm.
- ◆ Echocardiography shows abnormal motion in the left ventricular wall.
- ◆ MRI gives excellent visualization of the heart's apex.

## Treatment

Depending on the aneurysm's size and the complications, treatment may necessitate only routine medical examination to follow the patient's condition or aggressive measures for intractable ventricular arrhythmias, heart failure, and emboli.

Emergency treatment of ventricular arrhythmia includes antiarrhythmics I.V. or cardioversion. Preventive treatment continues with medications that decrease afterload to increase left ventricular function such as ACE inhibitors, anti-ischemic medications for angina, and anticoagulation if thrombus formation.

Emergency treatment for heart failure with pulmonary edema includes oxygen, cardiac glycosides I.V., furosemide I.V., morphine I.V., and, when necessary, nitroprusside I.V. and intubation. Maintenance therapy may include nitrates, prazosin, and oral hydralazine. Systemic embolization requires anticoagulation therapy or embolectomy. Refractory ventricular tachycardia, heart failure, recurrent arterial embolization, and persistent angina with coronary artery occlusion may necessitate surgery, of which the most effective procedure is aneurysmectomy with myocardial revascularization.

## Special Considerations

- ◆ If ventricular tachycardia occurs, administer a prescribed antiarrhythmic such as lidocaine. Monitor blood pressure and heart rate. If cardiac arrest develops, initiate CPR and call for assistance, resuscitative equipment, and medication.

- ◆ In a patient with heart failure, closely monitor vital signs, heart sounds, intake and output, fluid and electrolyte balances, and BUN and creatinine levels. Because of the threat of systemic embolization, frequently check peripheral pulses and the color and temperature of extremities. Be alert for sudden changes in sensorium that indicate cerebral embolization and for any signs that suggest renal failure or progressive MI.

If the patient is scheduled to undergo resection:

- ◆ Before surgery, explain expected postoperative care in the ICU (including use of such things as endotracheal [ET] tube, ventilator, hemodynamic monitoring, chest tubes, and drainage bottle).
- ◆ After surgery, monitor vital signs, intake and output, heart sounds, and pulmonary artery catheter. Watch for signs of infection, such as fever and drainage.

To prepare the patient for discharge:

- ◆ Teach how to check for pulse irregularity and rate changes. Encourage the patient to follow the prescribed medication regimen—even during the night—and to watch for adverse effects.
- ◆ Because arrhythmias can cause sudden death, refer the family to a community-based CPR training program.
- ◆ Provide psychological support for the patient and family.

## CARDIAC TAMPONADE

### Causes and Incidence

In cardiac tamponade, a rapid, unchecked rise in intrapericardial pressure impairs diastolic filling of the heart. The rise in pressure usually results from blood or fluid accumulation in the pericardial sac. If fluid accumulates rapidly, this condition requires emergency lifesaving measures to prevent death. A slow accumulation and rise in pressure, as in pericardial effusion associated with malignant tumors, may not produce immediate symptoms, because the fibrous wall of the pericardial sac can gradually stretch to accommodate as much as 1 to 2 L of fluid.

Increased intrapericardial pressure and cardiac tamponade may be idiopathic (Dressler syndrome) or may result from:

- ◆ effusion (in cancer, bacterial infections, tuberculosis, and, rarely, acute rheumatic fever)
- ◆ hemorrhage from trauma (such as gunshot or stab wounds of the chest and perforation by catheter during cardiac or central venous catheterization or postcardiac surgery)
- ◆ hemorrhage from nontraumatic causes (such as rupture of the heart or great vessels or anticoagulant therapy in a patient with pericarditis)
- ◆ acute MI
- ◆ end-stage lung cancer
- ◆ heart tumors
- ◆ radiation therapy
- ◆ hypothyroidism
- ◆ systemic lupus erythematosus
- ◆ uremia.

Cardiac tamponade occurs in 2 of every 10,000 people.

## Pathophysiology

Increased fluid in the pericardial space leads to increased pressure. The right ventricle is unable to expand adequately, and so pressure shifts the interventricular septum to the left. This decreases the left ventricle end-diastolic volume, resulting in decreased cardiac output and hypotension.

## Complications

- ◆ Cardiogenic shock
- ◆ Death

## Signs and Symptoms

Cardiac tamponade classically produces increased venous pressure with JVD, reduced arterial blood pressure, muffled heart sounds on auscultation, and pulsus paradoxus (an abnormal inspiratory drop in systemic blood pressure  $>15$  mm Hg). The absence of a preexisting pericardial friction rub may suggest an increase in fluid in the pericardial space. These classic symptoms represent failure of physiologic compensatory mechanisms to override the effects of rapidly rising pericardial pressure, which limits diastolic filling of the ventricles and reduces stroke volume to a critically low level. Generally, ventricular end-systolic volume may drop because of

inadequate preload. The increasing pericardial pressure is transmitted equally across the heart cavities, producing a matching rise in intracardiac pressure, especially atrial and end-diastolic ventricular pressures. Cardiac tamponade may also cause dyspnea, diaphoresis, pallor or cyanosis, anxiety, tachycardia, narrow pulse pressure, restlessness, and hepatomegaly, but the lung fields will be clear. The patient typically sits upright and leans forward.

## Diagnosis

- ◆ Chest X-ray shows slightly widened mediastinum and cardiomegaly.
- ◆ ECG is rarely diagnostic of tamponade but is useful in ruling out other cardiac disorders. It may reveal changes produced by acute pericarditis.
- ◆ Pulmonary artery catheterization detects increased right atrial pressure, right ventricular diastolic pressure, and CVP.
- ◆ Echocardiography, computed tomography scan, or MRI shows pericardial effusion with signs of right ventricular and atrial compression.

## Treatment

The goal of treatment is to relieve intrapericardial pressure and cardiac compression by removing accumulated blood or fluid. Pericardiocentesis (needle aspiration of the pericardial cavity) or surgical creation of an opening (pericardectomy or pericardial window) dramatically improves systemic arterial pressure and cardiac output with aspiration of as little as 25 mL of fluid. Such treatment necessitates continuous hemodynamic and ECG monitoring in the ICU. Trial volume loading with temporary I.V. normal saline solution and perhaps an inotropic drug, such as isoproterenol or dopamine, may be necessary in the hypotensive patient to maintain cardiac output. Although these drugs normally improve myocardial function, they may further compromise an ischemic myocardium after MI.

Depending on the cause of tamponade, additional treatment may include:

- ◆ in traumatic injury—blood transfusion or a thoracotomy to drain reaccumulating fluid or to repair bleeding sites
- ◆ in heparin-induced tamponade (not often used)—the heparin antagonist protamine sulfate
- ◆ in warfarin-induced tamponade—vitamin K.

Resection of a portion or all of the pericardium to allow full communication with the pleura may be needed if repeated pericardiocentesis fails to prevent recurrence.

## Special Considerations

If the patient needs pericardiocentesis:

- ◆ Explain the procedure to him. Keep a pericardial aspiration needle attached to a 50-mL syringe by a three-way stopcock, an ECG machine, and an emergency cart with a defibrillator at the bedside. Make sure the equipment is turned on and ready for immediate use. Position the patient at a 45- to 60-degree angle. Connect the precordial ECG lead to the hub of the aspiration needle with an alligator clamp and connecting wire, and assist with fluid aspiration. When the needle touches the myocardium, you'll see an ST-segment elevation or premature ventricular contractions.
- ◆ Monitor blood pressure and CVP during and after pericardiocentesis. Infuse I.V. solutions, as prescribed, to maintain blood pressure. Watch for a decrease in CVP and a concomitant rise in blood pressure, which indicate relief of cardiac compression.
- ◆ Watch for complications of pericardiocentesis, such as ventricular fibrillation, vasovagal response, or coronary artery or cardiac chamber puncture. Closely monitor ECG changes, blood pressure, pulse rate, LOC, and urine output.

If the patient needs thoracotomy:

- ◆ Explain the procedure to him. Tell the patient what to expect postoperatively (chest tubes, drainage bottles, and oxygen administration). Teach the patient how to turn, deep breathe, and cough.
- ◆ Give antibiotics, protamine sulfate, or vitamin K, as ordered.
- ◆ Postoperatively, monitor critical parameters, such as vital signs and ABG values, and assess heart and breath sounds. Give pain medication as ordered. Maintain the chest drainage system and be alert for complications, such as hemorrhage and arrhythmias.



**PREVENTION** Instruct patient to practice heart-healthy living, with a heart-healthy diet, stress reduction, regular exercise and preventive care,

*maintenance of a healthy weight, smoking cessation, and abstinence from alcohol.*

## CARDIAC ARRHYTHMIAS

### Causes and Incidence

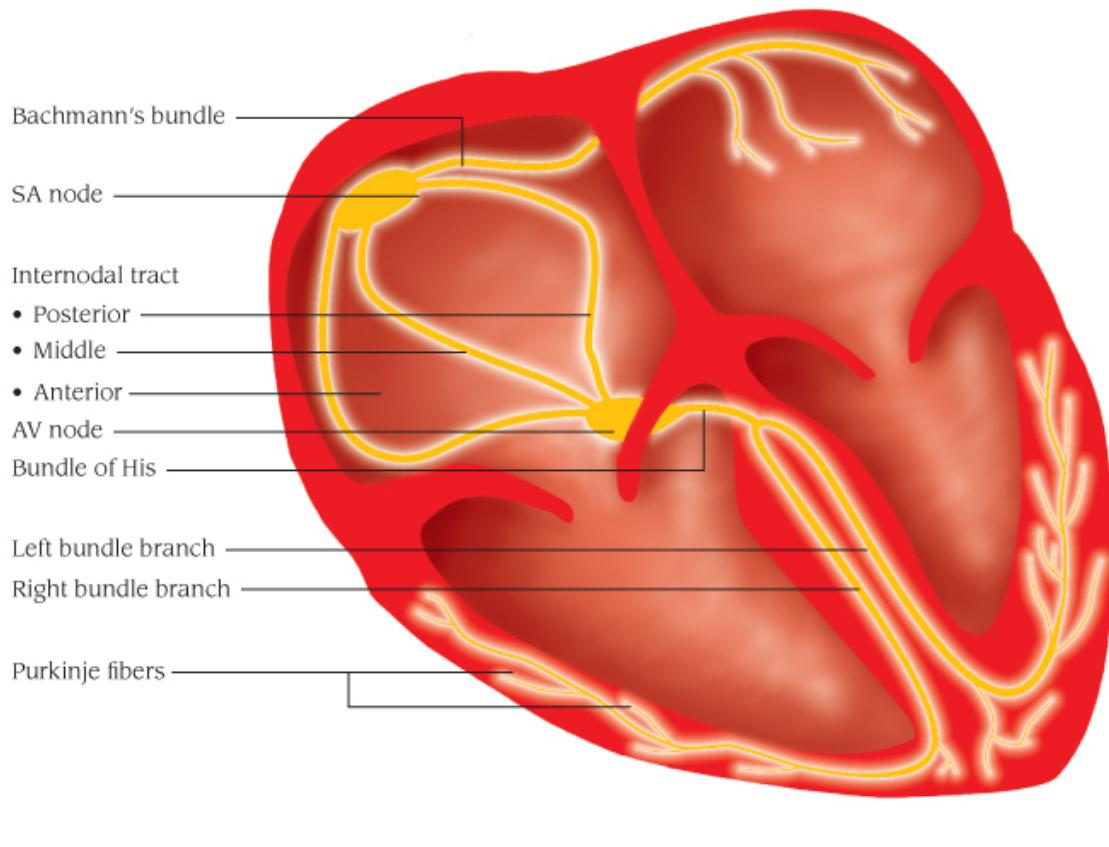
Arrhythmias may be congenital or may result from one of several factors, including myocardial ischemia, MI, or organic heart disease. Drug ingestion (cocaine, amphetamines, caffeine, BBs, psychotropics, sympathomimetics), drug toxicity, or degeneration of the conductive tissue necessary to maintain normal heart rhythm (sick sinus syndrome) can sometimes precipitate arrhythmias. People with imbalances of blood chemistries or those with a history of cardiac conditions (CAD or heart valve disorders) are at higher risk for developing arrhythmias.

### Pathophysiology

In cardiac arrhythmias (sometimes called *cardiac dysrhythmias*), abnormal electrical conduction or automaticity changes heart rate and rhythm. (See *Normal cardiac conduction*, page 61.) Arrhythmias vary in severity, from those that are mild, asymptomatic, and require no treatment (such as sinus arrhythmia, in which heart rate increases and decreases with respiration) to catastrophic ventricular fibrillation, which necessitates immediate resuscitation. Arrhythmias are generally classified according to their origin (ventricular or supraventricular). Their effect on cardiac output and blood pressure, partially influenced by the site of origin, determines their clinical significance. (See *Types of cardiac arrhythmias*, pages 65 to 71.)

### Normal Cardiac Conduction

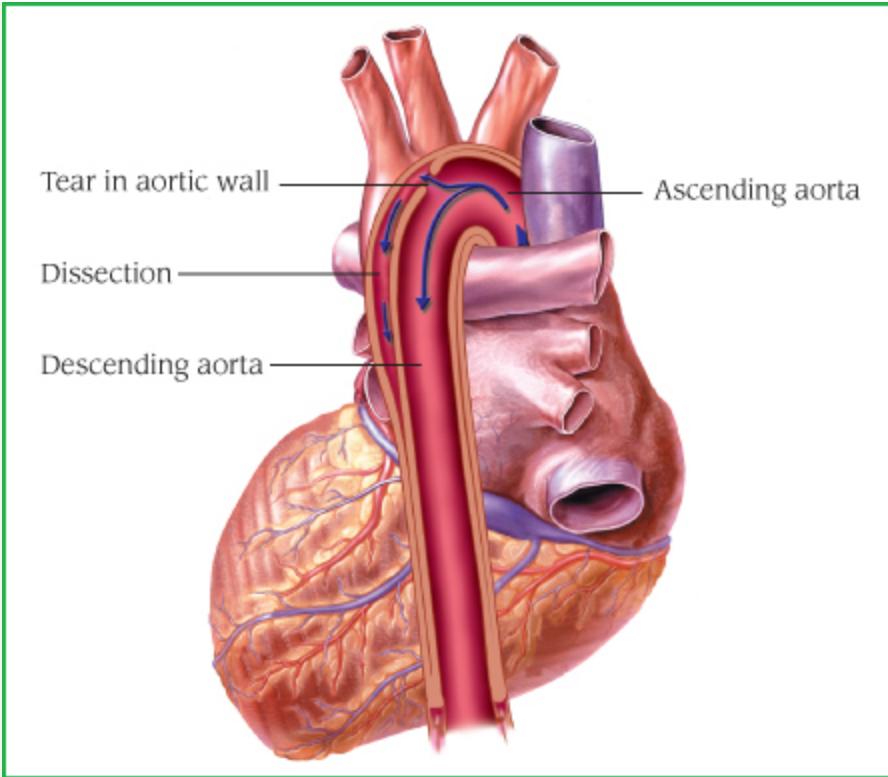
The conduction system of the heart begins with the heart's pacemaker: the sinoatrial (SA) node. When an impulse leaves the SA node, it travels through the atria along the Bachmann bundle and the internodal tracts on its way to the atrioventricular (AV) node. After the impulse passes through the AV node, it travels to the ventricles, first down the bundle of His, then along the bundle branches and, finally, down the Purkinje fibers.



## Types of Aortic Aneurysms

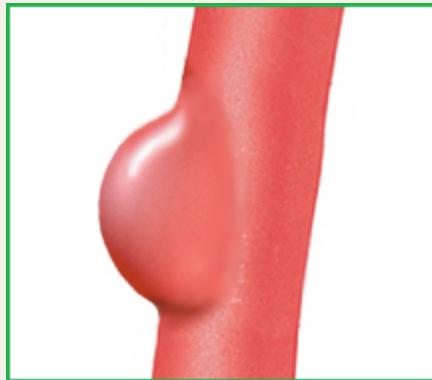
### Dissecting Aneurysm

A hemorrhagic separation of the medial layer of the vessel wall, which creates a false lumen



### Saccular Aneurysm

Unilateral pouchlike bulge with a narrow neck



### Fusiform Aneurysm

A spindle-shaped bulge encompassing the vessel's entire diameter



- ◆ syphilis, usually of the ascending aorta (uncommon because of antibiotics)
- ◆ hypertension (in dissecting aneurysm).

### False Aneurysm

A pulsating hematoma resulting from trauma; usually seen in the femoral artery after catheterization

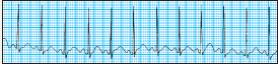


### Types of Cardiac Arrhythmias

This chart reviews many common cardiac arrhythmias and outlines their features, causes, and treatments. Use a normal electrocardiogram strip, if available, to compare normal cardiac rhythm configurations with the rhythm strips below. Characteristics of normal rhythm include:

- ◆ ventricular and atrial rates of 60 to 100 beats/minute
- ◆ regular and uniform QRS complexes and P waves

- ◆ PR interval of 0.12 to 0.2 second
- ◆ QRS duration <0.12 second
- ◆ identical atrial and ventricular rates, with constant PR interval.

| <b>Arrhythmia and features</b>                        | <b>Causes</b>  | <b>Treatment</b>  |
|---|--|---|
| <b>Sinus arrhythmia</b>                               |  <ul style="list-style-type: none"> <li>◆ Irregular atrial and ventricular rhythms</li> <li>◆ Normal P wave preceding each QRS complex</li> </ul>   | <ul style="list-style-type: none"> <li>◆ A normal variation of normal sinus rhythm in athletes, children, and elderly people</li> <li>◆ Also seen in digoxin toxicity and inferior wall myocardial infarction (MI)</li> </ul> <p>◆ Atropine if rate decreases below 40 beats/minute and patient is symptomatic (e.g., has hypotension)</p>  |
| <b>Sinus tachycardia</b>                              |  <ul style="list-style-type: none"> <li>◆ Atrial and ventricular rhythms regular</li> <li>◆ Rate &gt; 100 beats/minute; rarely, &gt;160 beats/minute</li> <li>◆ Normal P wave preceding each QRS complex</li> </ul> | <ul style="list-style-type: none"> <li>◆ Normal physiologic response to fever, exercise, anxiety, pain, dehydration; may also accompany shock, left-sided heart failure, cardiac tamponade, hyperthyroidism, anemia, hypovolemia, pulmonary embolism, and anterior wall MI</li> <li>◆ May also occur with atropine, epinephrine, isoproterenol, quinidine, caffeine, alcohol, and nicotine use</li> </ul> <p>◆ Correction of underlying cause</p> <p>◆ Beta-adrenergic blockers or calcium channel blockers for symptomatic patients</p>  |
| <b>Sinus bradycardia</b>                              |  <ul style="list-style-type: none"> <li>◆ Regular atrial and ventricular rhythms</li> <li>◆ Rate &lt; 60 beats/minute</li> <li>◆ Normal P wave preceding each QRS complex</li> </ul>                              | <ul style="list-style-type: none"> <li>◆ Normal in well-conditioned heart, as in an athlete</li> <li>◆ Increased intracranial pressure; increased vagal tone due to straining during defecation, vomiting, intubation, mechanical ventilation; sick sinus syndrome; hypothyroidism; inferior wall MI</li> <li>◆ May also occur with anticholinesterase, beta-adrenergic blocker, digoxin, and morphine use</li> </ul> <p>◆ For low cardiac output, dizziness, weakness, altered level of consciousness, or low blood pressure: follow advanced cardiac life support (ACLS) protocol for administration of atropine</p> <p>◆ Temporary pacemaker; may need to be evaluated for permanent pacemaker at a later time</p> |
| <b>Sinoatrial (SA) arrest or block (sinus arrest)</b> |  |   |

| <b>Arrhythmia and features</b>  | <b>Causes</b>   | <b>Treatment</b>   |
|---|---|--|
|  | <ul style="list-style-type: none"> <li>◆ Atrial and ventricular rhythms normal except for missing complex</li> <li>◆ Normal P wave preceding each QRS complex</li> <li>◆ Pause not equal to a multiple of the previous sinus rhythm</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Acute infection</li> <li>◆ Coronary artery disease, degenerative heart disease, and acute inferior wall MI</li> <li>◆ Vagal stimulation, Valsalva maneuver, or carotid sinus massage</li> <li>◆ Digoxin, quinidine, or salicylate toxicity</li> <li>◆ Pesticide poisoning</li> <li>◆ Pharyngeal irritation caused by endotracheal (ET) intubation</li> <li>◆ Sick sinus syndrome</li> </ul> |
| <b>Wandering atrial pacemaker</b>   |   |  |
|  | <ul style="list-style-type: none"> <li>◆ Atrial and ventricular rhythms vary slightly</li> <li>◆ Irregular PR interval</li> <li>◆ P waves irregular with changing configuration, indicating that they aren't all from SA node or single atrial focus; may appear after the QRS complex</li> <li>◆ QRS complexes uniform in shape but irregular in rhythm</li> </ul> | <ul style="list-style-type: none"> <li>◆ Rheumatic carditis due to inflammation involving the SA node</li> <li>◆ Digoxin toxicity</li> <li>◆ Sick sinus syndrome</li> </ul>  |
| <b>Premature atrial contraction (PAC)</b>   |   |  |

| <b>Arrhythmia and features</b>  | <b>Causes</b>  | <b>Treatment</b>   |
|---|--|--|
|  <p>Premature, abnormal-looking P waves that</p> <ul style="list-style-type: none"> <li>◆ differ in configuration from normal P waves</li> <li>◆ QRS complexes after P waves, except in very early or blocked PACs</li> <li>◆ P wave often buried in the preceding T wave or identified in the preceding T wave</li> </ul> | <ul style="list-style-type: none"> <li>◆ Coronary or valvular heart disease, atrial ischemia, coronary atherosclerosis, heart failure, acute respiratory failure, chronic obstructive pulmonary disease (COPD), electrolyte imbalance, and hypoxia</li> <li>◆ Digoxin toxicity; use of aminophylline, adrenergics, or caffeine</li> <li>◆ Anxiety</li> </ul> | <ul style="list-style-type: none"> <li>◆ Usually no treatment needed</li> <li>◆ Treatment of underlying cause if patient is symptomatic</li> </ul> |

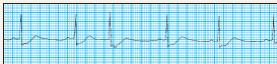
### **Paroxysmal supraventricular tachycardia**

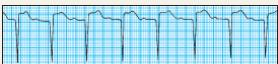
| <b>Arrhythmia and features</b>   | <b>Causes</b>   | <b>Treatment</b>  |
|--|---|---|
|  <ul style="list-style-type: none"> <li>◆ Atrial and ventricular rhythms regular</li> <li>◆ Heart rate &gt;160 beats/minute; rarely exceeds 250 beats/minute</li> <li>◆ P waves regular but aberrant; difficult to differentiate from preceding T wave</li> <li>◆ P wave preceding each QRS complex</li> <li>◆ Sudden onset and termination of arrhythmia</li> <li>◆ When a normal P wave is present, it's called <i>paroxysmal atrial tachycardia</i>; when a normal P wave isn't present, it's called <i>paroxysmal junctional tachycardia</i></li> </ul> | <ul style="list-style-type: none"> <li>◆ Intrinsic abnormality of atrioventricular (AV) conduction system</li> <li>◆ Physical or psychological stress, hypoxia, hypokalemia, cardiomyopathy, congenital heart disease, MI, valvular disease, Wolff–Parkinson–White syndrome, cor pulmonale, hyperthyroidism, and systemic hypertension</li> <li>◆ Digoxin toxicity; use of caffeine, marijuana, or central nervous system stimulants</li> </ul> | <ul style="list-style-type: none"> <li>◆ If patient is unstable, prepare for immediate cardioversion</li> <li>◆ If patient is stable, vagal stimulation, Valsalva maneuver, or carotid sinus massage</li> <li>◆ Adenosine by rapid I.V. bolus injection to rapidly convert arrhythmia</li> <li>◆ If patient has a normal ejection fraction, consider calcium channel blockers, beta-adrenergic blockers, or amiodarone</li> <li>◆ If patient has an ejection fraction &lt;40%, consider amiodarone</li> </ul> |

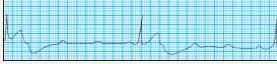
### **Atrial flutter**

| <b>Arrhythmia and features</b>   | <b>Causes</b>  | <b>Treatment</b>  |
|--|--|---|
| <p></p> <ul style="list-style-type: none"> <li>◆ Atrial rhythm regular; rate, 250 to 400 beats/minute</li> <li>◆ Ventricular rate variable, depending on degree of AV block (usually 60 to 100 beats/minute)</li> <li>◆ Sawtooth P-wave configuration possible (F waves)</li> <li>◆ QRS complexes uniform in shape but often irregular in rate</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Heart failure, tricuspid or mitral valve disease, pulmonary embolism, cor pulmonale, inferior wall MI, and carditis</li> <li>◆ Digoxin toxicity</li> </ul>  | <ul style="list-style-type: none"> <li>◆ If patient is unstable with a ventricular rate &gt;150 beats/minute, prepare for immediate cardioversion</li> <li>◆ If patient is stable, drug therapy may include calcium channel blockers, beta-adrenergic blockers, or antiarrhythmics</li> <li>◆ Anticoagulation therapy may be necessary</li> <li>◆ Catheter ablation using radiofrequency energy to eliminate cardiac tissue causing the rapid heartbeat</li> </ul>  |
| <p><b>Atrial fibrillation</b></p> <p></p> <ul style="list-style-type: none"> <li>◆ Atrial rhythm grossly irregular; rate &gt;400 beats/minute</li> <li>◆ Ventricular rhythm grossly irregular</li> <li>◆ QRS complexes of uniform configuration and duration</li> <li>◆ PR interval indiscernible</li> <li>◆ No P waves, or P waves that appear as erratic, irregular, baseline fibrillatory waves</li> </ul> | <ul style="list-style-type: none"> <li>◆ Heart failure, COPD, thyrotoxicosis, constrictive pericarditis, ischemic heart disease, sepsis, pulmonary embolus, rheumatic heart disease, hypertension, mitral stenosis, and atrial irritation; complication of coronary bypass or valve replacement surgery</li> </ul> | <ul style="list-style-type: none"> <li>◆ If patient is unstable with a ventricular rate &gt;150 beats/minute, prepare for immediate cardioversion</li> <li>◆ If patient is stable, drug therapy may include calcium channel blockers, beta-adrenergic blockers, digoxin, procainamide, quinidine, ibutilide, or amiodarone</li> <li>◆ Consider anticoagulation to prevent emboli</li> <li>◆ Dual-chamber atrial pacing, implantable atrial pacemaker, or surgical maze procedure may also be used</li> <li>◆ Catheter ablation</li> </ul> |
| <p><b>Junctional rhythm</b></p>  |  |   |

| <b>Arrhythmia and features</b>  | <b>Causes</b>   | <b>Treatment</b>   |
|---|---|--|
|  <ul style="list-style-type: none"> <li>◆ Atrial and ventricular rhythms regular</li> <li>◆ Atrial rate 40 to 60 beats/minute</li> <li>◆ Ventricular rate usually 40 to 60 beats/minute (60 to 100 beats/minute is accelerated junctional rhythm)</li> <li>◆ P waves preceding, hidden within (absent), or after QRS complex; usually inverted if visible</li> <li>◆ PR interval (when present) &lt;0.12 second</li> <li>◆ QRS complex configuration and duration normal, except in aberrant conduction</li> </ul> | <ul style="list-style-type: none"> <li>◆ Inferior wall MI or ischemia, hypoxia, vagal stimulation, and sick sinus syndrome</li> <li>◆ Acute rheumatic fever</li> <li>◆ Valve surgery</li> <li>◆ Digoxin toxicity</li> </ul> | <ul style="list-style-type: none"> <li>◆ Correction of underlying cause</li> <li>◆ Atropine for symptomatic slow rate</li> <li>◆ Pacemaker insertion if patient is refractory to drugs</li> <li>◆ Discontinuation of digoxin if appropriate</li> </ul> |
| <b>Premature junctional contractions</b>  |   |  |

| <b>Arrhythmia and features</b>  | <b>Causes</b>   | <b>Treatment</b>  |
|---|---|---|
|    | <ul style="list-style-type: none"> <li>◆ MI or ischemia</li> <li>◆ Digoxin toxicity and excessive caffeine or amphetamine use</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Correction of underlying cause</li> <li>◆ Discontinuation of digoxin if appropriate</li> </ul>   |
| <ul style="list-style-type: none"> <li>◆ Atrial and ventricular rhythms irregular</li> <li>◆ P waves inverted; may precede, be hidden within, or follow QRS complex</li> <li>◆ PR interval &lt;0.12 second if P wave precedes QRS complex</li> <li>◆ QRS complex configuration and duration normal</li> </ul>   |   |   |
| <b>Junctional tachycardia</b>   |   |   |
|    | <ul style="list-style-type: none"> <li>◆ Myocarditis, cardiomyopathy, inferior wall MI or ischemia, and acute rheumatic fever; complication of valve replacement surgery</li> <li>◆ Digoxin toxicity</li> </ul> | <ul style="list-style-type: none"> <li>◆ Cardioversion if ventricular rate is &gt;150 beats/minute or if patient is symptomatic</li> <li>◆ Amiodarone, beta-adrenergic blockers, or calcium channel blockers if patient is stable</li> <li>◆ Discontinuation of digoxin if appropriate</li> </ul> |
| <ul style="list-style-type: none"> <li>◆ Atrial rate &gt;100 beats/minute; however, P wave may be absent, hidden in QRS complex, or preceding T wave</li> <li>◆ Ventricular rate &gt;100 beats/minute</li> <li>◆ P wave inverted</li> <li>◆ QRS complex configuration and duration normal</li> <li>◆ Onset of rhythm often sudden, occurring in bursts</li> </ul> |   |   |
| <b>First-degree AV block</b>  |   |   |

| <b>Arrhythmia and features</b>  | <b>Causes</b>  | <b>Treatment</b>   |
|---|--|--|
|  <ul style="list-style-type: none"> <li>◆ Atrial and ventricular rhythms regular</li> <li>◆ PR interval &gt;0.20 second</li> <li>◆ P wave preceding each QRS complex</li> <li>◆ QRS complex normal</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Inferior wall MI or ischemia or infarction, hypothyroidism, hypokalemia, and hyperkalemia</li> <li>◆ Digoxin toxicity; use of quinidine, procainamide, beta-adrenergic blockers, calcium channel blockers, or amiodarone</li> </ul> | <ul style="list-style-type: none"> <li>◆ Correction of underlying cause</li> <li>◆ No treatment is typically required</li> </ul>   |
| <b>Second-degree AV block Mobitz I (Wenckebach)</b>   |  |  |
|  <ul style="list-style-type: none"> <li>◆ Atrial rhythm regular</li> <li>◆ Ventricular rhythm irregular</li> <li>◆ Atrial rate exceeds ventricular rate</li> <li>◆ PR interval progressively, but only slightly, longer with each cycle until QRS complex disappears (dropped beat); PR interval shorter after dropped beat</li> </ul> | <ul style="list-style-type: none"> <li>◆ Inferior wall MI, cardiac surgery, acute rheumatic fever, and vagal stimulation</li> <li>◆ Digoxin toxicity; use of propranolol, quinidine, or procainamide</li> </ul>  | <ul style="list-style-type: none"> <li>◆ No specific treatment is required if asymptomatic</li> <li>◆ Treatment of underlying cause</li> <li>◆ Atropine or temporary pacemaker for symptomatic bradycardia</li> <li>◆ Discontinuation of digoxin if appropriate</li> </ul> |
| <b>Second-degree AV block Mobitz II</b>   |  |  |

| <b>Arrhythmia and features</b>  | <b>Causes</b>  | <b>Treatment</b>  |
|---|--|---|
|  | <ul style="list-style-type: none"> <li>◆ Severe coronary artery disease, anterior wall MI, and acute myocarditis</li> <li>◆ Digoxin toxicity</li> </ul> <ul style="list-style-type: none"> <li>◆ Atrial rhythm regular</li> <li>◆ Ventricular rhythm regular or irregular, with varying degree of block</li> <li>◆ P-P interval constant</li> <li>◆ QRS complexes periodically absent</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Atropine, for symptomatic bradycardia</li> <li>◆ Dopamine for hypotension</li> <li>◆ Dobutamine for heart failure symptoms</li> <li>◆ Temporary or permanent pacemaker for symptomatic bradycardia</li> <li>◆ Discontinuation of digoxin if appropriate</li> </ul> |
| <b>Third-degree AV block (complete heart block)</b>                               |  |   |
|  | <ul style="list-style-type: none"> <li>◆ Inferior or anterior wall MI, congenital abnormality, rheumatic fever, hypoxia, postoperative complication of mitral valve replacement, Lev disease (fibrosis and calcification that spreads from cardiac structures to the conductive tissue), and Lenegre disease (conductive tissue fibrosis)</li> <li>◆ Digoxin toxicity</li> </ul> <ul style="list-style-type: none"> <li>◆ Atrial rhythm regular</li> <li>◆ Ventricular rhythm regular and rate slower than atrial rate</li> <li>◆ No relation between P waves and QRS complexes</li> <li>◆ No constant PR interval</li> <li>◆ QRS interval normal (nodal pacemaker) or wide and bizarre (ventricular pacemaker)</li> </ul> | <ul style="list-style-type: none"> <li>◆ Atropine, for symptomatic bradycardia</li> <li>◆ Dopamine for hypotension</li> <li>◆ Dobutamine for heart failure symptoms</li> <li>◆ Temporary or permanent pacemaker for symptomatic bradycardia</li> </ul>  |
| <b>Premature ventricular contraction (PVC)</b>                                    |  |   |

| <b>Arrhythmia and features</b>   | <b>Causes</b>  | <b>Treatment</b>   |
|--|--|--|
|  <ul style="list-style-type: none"> <li>◆ Atrial rhythm regular</li> <li>◆ Ventricular rhythm irregular</li> <li>◆ QRS complex</li> <li>◆ premature, usually followed by a complete compensatory pause</li> <li>◆ QRS complex wide and distorted, usually &gt;0.14 second</li> <li>◆ Premature QRS complexes occurring singly, in pairs, or in threes; alternating with normal beats; focus from one or more sites</li> <li>◆ Ominous when clustered, multifocal, with R wave on T pattern</li> </ul> | <ul style="list-style-type: none"> <li>◆ Heart failure; old or acute myocardial ischemia, infarction, or contusion; myocardial irritation by ventricular catheter such as a pacemaker; hypercapnia; hypokalemia; and hypocalcemia</li> <li>◆ Drug toxicity (cardiac glycosides, aminophylline, tricyclic antidepressants, beta-adrenergics [isoproterenol or dopamine])</li> <li>◆ Caffeine, tobacco, or alcohol use</li> <li>◆ Psychological stress, anxiety, pain, exercise</li> </ul> | <ul style="list-style-type: none"> <li>◆ Treatment of underlying cause</li> <li>◆ Discontinuation of drug causing toxicity</li> <li>◆ Correction of electrolyte imbalances</li> <li>◆ Beta-blockers, calcium channel blockers, or antiarrhythmics may be used to treat symptoms</li> </ul> |
| <b>Ventricular tachycardia (VT)</b>  |  |  |

| <b>Arrhythmia and features</b>  | <b>Causes</b>   | <b>Treatment</b>  |
|---|---|---|
|  <ul style="list-style-type: none"> <li>◆ Ventricular rate 140 to 220 beats/minute, regular or irregular</li> <li>◆ QRS complexes wide, bizarre, and independent of P waves</li> <li>◆ P waves not discernible</li> <li>◆ May start and stop suddenly</li> </ul> | <ul style="list-style-type: none"> <li>◆ Myocardial ischemia, infarction, or aneurysm; coronary artery disease; rheumatic heart disease; mitral valve prolapse; heart failure; cardiomyopathy; ventricular catheters; hypokalemia; hypercalcemia; and pulmonary embolism</li> <li>◆ Digoxin, procainamide, epinephrine, or quinidine toxicity</li> <li>◆ Anxiety</li> </ul> | <ul style="list-style-type: none"> <li>◆ Pulseless: Initiate cardiopulmonary resuscitation (CPR); follow ACLS protocol for defibrillation, ET intubation, and administration of epinephrine by amiodarone or lidocaine; if ineffective, consider magnesium sulfate</li> <li>◆ With pulse: If hemodynamically stable, follow ACLS protocol for administration of amiodarone; if ineffective, initiate synchronized cardioversion</li> <li>◆ If polymorphic VT, consult an expert in arrhythmia management</li> </ul> |
| <b>Ventricular fibrillation</b>  <ul style="list-style-type: none"> <li>◆ Ventricular rhythm and rate rapid and chaotic</li> <li>◆ QRS complexes wide and irregular; no visible P waves</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Myocardial ischemia or infarction, R-on-T phenomenon, untreated ventricular tachycardia, hypokalemia, hyperkalemia, hypercalcemia, alkalosis, electric shock, and hypothermia</li> <li>◆ Digoxin, epinephrine, or quinidine toxicity</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Pulseless: Start CPR; follow ACLS protocol for defibrillation, ET intubation, and administration of epinephrine, lidocaine, or amiodarone; if ineffective, consider magnesium sulfate</li> </ul>   |
| <b>Asystole</b>  <ul style="list-style-type: none"> <li>◆ No atrial or ventricular rate or rhythm</li> <li>◆ No discernible P waves, QRS complexes, or T waves</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Myocardial ischemia or infarction, aortic valve disease, heart failure, hypoxemia, hypokalemia, severe acidosis, electric shock, ventricular arrhythmias, AV block, pulmonary embolism, heart rupture, cardiac tamponade, hyperkalemia, and electromechanical dissociation</li> <li>◆ Cocaine overdose</li> </ul>                  | <ul style="list-style-type: none"> <li>◆ Start CPR; follow ACLS protocol for ET intubation, transcutaneous pacing, and administration of epinephrine</li> </ul>   |

## **Complications**

- ◆ Impaired cardiac output

## **Signs and Symptoms**

Signs and symptoms of cardiac arrhythmias include palpitations, fainting, light-headedness, dizziness, chest pain, shortness of breath, changes in pulse patterns, paleness, and the temporary absence of breathing. However, the patient with a cardiac arrhythmia may be asymptomatic until the development of sudden cardiac arrest.

## **Diagnosis**

Diagnosis is made by tests that reveal the arrhythmia, such as 12-lead ECG. Ambulatory cardiac monitoring (Holter monitoring), echocardiography, electrophysiology studies, and coronary angiography may also confirm or rule out suspected causes of arrhythmias and help determine treatment.

## **Special Considerations**

- ◆ Assess an unmonitored patient for rhythm disturbances.
- ◆ If the patient's pulse is abnormally rapid, slow, or irregular, watch for signs of hypoperfusion, such as altered LOC, hypotension, and diminished urine output.
- ◆ Document arrhythmias in a monitored patient, and assess for possible causes and effects.
- ◆ When life-threatening arrhythmias develop, rapidly assess LOC, respirations, and pulse rate.
- ◆ Initiate CPR if indicated.
- ◆ Evaluate the patient for altered cardiac output resulting from arrhythmias.
- ◆ Administer medications as ordered and prepare to assist with medical procedures (e.g., cardioversion), if indicated.
- ◆ Monitor patient for predisposing factors—such as fluid and electrolyte imbalance—and signs of drug toxicity, especially with digoxin. If you suspect drug toxicity, report such signs to the practitioner immediately and withhold the next dose.
- ◆ To prevent arrhythmias in a postoperative cardiac patient, provide adequate oxygen and reduce the heart's workload while carefully

- maintaining metabolic, neurologic, respiratory, and hemodynamic status.
- ◆ Consider sedation for transcutaneous pacing if appropriate.
  - ◆ To avoid temporary pacemaker malfunction, install a fresh battery before each insertion. Carefully secure the external catheter wires and the pacemaker box. Assess the threshold daily. Watch closely for premature contractions, a sign of myocardial irritation.
  - ◆ To avert permanent pacemaker malfunction, restrict the patient's activity after insertion as ordered. Monitor the pulse rate regularly and watch for signs of decreased cardiac output.
  - ◆ If the patient has a permanent pacemaker, warn about environmental hazards, as indicated by the pacemaker manufacturer. Although hazards may not present a problem, in doubtful situations, 24-hour Holter monitoring may be helpful. Tell the patient to report light-headedness or syncope, and stress the importance of regular checkups.
  - ◆ Compare the patient's cardiac status (pulse, blood pressure, and cardiac output) with the cardiac rhythm before and after treatments.



## PREVENTION

- *Maintain adequate oxygenation.*
- *Maintain normal fluid, acid-base, and electrolyte (especially potassium, magnesium, and calcium) balance.*
- *Maintain normal drug levels.*

# Vascular Disorders

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## THORACIC AORTIC ANEURYSM

### Causes and Incidence

Thoracic aortic aneurysm is an abnormal widening of the ascending, transverse, or descending part of the aorta. Aneurysm of the ascending aorta is the most common type and has the highest mortality. Aneurysms may be *dissecting*, a hemorrhagic separation in the aortic wall, usually within the medial layer; *saccular*, an outpouching of the arterial wall, with a narrow neck; or *fusiform*, a spindle-shaped enlargement encompassing the entire aortic circumference. (See *Types of aortic aneurysms*, page 64.) Some

aneurysms progress to serious and, eventually, lethal complications, such as rupture of an untreated thoracic dissecting aneurysm into the pericardium, with resulting tamponade.

Thoracic aortic aneurysms commonly result from atherosclerosis, which weakens the aortic wall and gradually distends the lumen. An intimal tear in the ascending aorta initiates dissecting aneurysm in about 65% of patients. Regardless of causation, these aneurysms affect 6 out of every 100,000 people.



**ELDER TIP** *Ascending aortic aneurysms, the most common type, are usually seen in hypertensive men younger than 60 years old.*

*Descending aortic aneurysms, usually found just below the origin of the subclavian artery, are most common in elderly men who are hypertensive.*

Descending aortic aneurysms are also seen in younger patients with a history of traumatic chest injury less often in those with infection. Transverse aortic aneurysms are the least common type.

Other causes include:

- ◆ fungal infection (mycotic aneurysms) of the aortic arch and descending segments
- ◆ congenital disorders, such as coarctation of the aorta and Marfan syndrome
- ◆ trauma, usually of the descending thoracic aorta, from an accident that shears the aorta transversely (acceleration–deceleration injuries)

## Pathophysiology

Most aneurysms result from changes to the vascular wall of arteries caused by failure of important proteins, elastin and collagen, leading to dilation. This eventually affects the structural stability and strength of the vessel. The location of the aneurysm, its diameter, the cause, and its morphology all assist in determining rate of expansion per year. Saccular aneurysms are thought to rupture more readily than other types, such as fusiform.

## Complications

- ◆ Rupture into pericardium
- ◆ Cardiac tamponade

## Signs and Symptoms

The most common symptom of thoracic aortic aneurysm is pain. With ascending aneurysm, the pain is described as severe, boring, and ripping and extends to the neck, shoulders, lower back, or abdomen but seldom radiates to the jaw and arms. Pain is more severe on the right side.

Other signs of ascending aneurysm may include bradycardia, aortic insufficiency, pericardial friction rub caused by a hemopericardium, unequal intensities of the right carotid and left radial pulses, and a difference in blood pressure between the right and the left arms. These signs are absent in descending aneurysm. If dissection involves the carotids, an abrupt onset of neurologic deficits may occur.

With descending aneurysm, pain usually starts suddenly between the shoulder blades and may radiate to the chest; it's described as sharp and tearing. Transverse aneurysm causes a sudden, sharp, tearing pain radiating to the shoulders. It may also cause hoarseness, dyspnea, dysphagia, and a dry cough because of compression of surrounding structures in this area. (See *Clinical characteristics of thoracic dissection*, page 72.)

## Clinical Characteristics of Thoracic Dissection

| Ascending aorta   | Descending aorta   | Transverse aorta   |
|---|--|--|
| <b>Character of pain</b>  |  |  |
| Severe, boring, ripping; extending to neck, shoulders, lower back, or abdomen (rarely to jaw and arms); more severe on right side   | Sudden onset; sharp, tearing; usually between the shoulder blades; may radiate to the chest; most diagnostic feature                     | Sudden onset; sharp, boring, tearing; radiates to shoulders  |
| <b>Other symptoms and effects</b>   |  |  |
| If dissection involves carotids, abrupt onset of neurologic deficit (usually intermittent); bradycardia, aortic insufficiency, and hemopericardium detected by pericardial friction rub; unequal intensity of right and left carotid pulses and radial pulses; difference in blood pressure, especially systolic, between right and left arms | Aortic insufficiency without murmur, hemopericardium, or pleural friction rub; carotid and radial pulses and blood pressure in both arms | Hoarseness, dyspnea, pain, dysphagia, and dry cough resulting from compression of surrounding structures |
| <b>Diagnostic features</b>  |  |  |
| <b>Chest X-ray</b>  |  |  |

| <b>Ascending aorta</b>  | <b>Descending aorta</b>  | <b>Transverse aorta</b>   |
|---|--|---|
| Best diagnostic tool; shows widening of mediastinum, enlargement of ascending aorta   | Shows widening of mediastinum, descending aorta larger than ascending  | Shows widening of mediastinum, descending aorta larger than ascending, widened transverse arch  |
| <b>Computerized tomography</b>  |  |   |
| Shows false lumen; narrowing of lumen of aorta in ascending section. Also good for visualizing anatomic details to help determine the extent of the aneurysm, and to assist in preparing for repair                                       |  |   |
|   | Shows false lumen; narrowing of lumen of aorta in descending section. Also good for visualizing anatomic details to help determine the extent of the aneurysm, and to assist in preparing for repair | Shows false lumen, narrowing of lumen of aorta in transverse arch. Also good for visualizing anatomic details to help determine the extent of the aneurysm, and to assist in preparing for repair |
| <b>Treatment</b>  |  |   |
| This is a medical emergency requiring immediate, aggressive treatment to reduce blood pressure (usually with labetalol or verapamil). Nitroprusside may be required if there is persistent hypertension. Surgical repair is also required | Surgical repair is required but less urgent than for the ascending dissection  | Immediate surgical repair (mortality as high as 50%) and control of hypertension are required   |

## Diagnosis

Diagnosis relies on patient history, clinical features, and appropriate tests. In an asymptomatic patient, diagnosis often occurs accidentally when chest X-rays show widening of the aorta. Other tests help confirm aneurysm:

- ◆ Aortography can show the lumen of the aneurysm, its size and location, and the false lumen in dissecting aneurysm.
- ◆ ECG helps distinguish thoracic aneurysm from MI.
- ◆ Echocardiography may help identify dissecting aneurysm of the aortic root.
- ◆ Hemoglobin levels may be normal or low, due to blood loss from a leaking aneurysm.
- ◆ Computed tomography scan is considered the gold standard and can confirm and locate the aneurysm and may be used to monitor its progression.

- ◆ MRI may aid diagnosis.
- ◆ Transesophageal echocardiography is used to diagnose and size an aneurysm in either the ascending or the descending aorta.
- ◆ Ultrasound is frequently used for screening purposes in high-risk patients.

## Treatment

Dissecting aortic aneurysm is an emergency that requires prompt surgery and stabilizing measures: antihypertensives such as nitroprusside; negative inotropic agents that decrease contractility force such as propranolol; oxygen for respiratory distress; opioids for pain; I.V. fluids and, possibly, whole blood transfusions.

Surgery consists of resecting the aneurysm, restoring normal blood flow through a graft replacement, and, with aortic valve insufficiency, replacing the aortic valve. Groin catheter placement may be used for aortic stenting. This procedure, which may be used for aneurysms of the descending aorta, eliminates the need for a chest incision.

Postoperative measures include careful monitoring and continuous assessment in the ICU, antibiotics, placement of ET and chest tubes, ECG monitoring, and pulmonary artery catheterization.

Long-term management includes treatment of underlying conditions, such as heart disease and diabetes.

## Special Considerations

- ◆ Monitor blood pressure, PAWP, and CVP. Assess pain; breathing; and carotid, radial, and femoral pulses.
- ◆ Make sure laboratory tests include complete blood count, differential, electrolyte levels, type and crossmatching for whole blood, ABG studies, and urinalysis.
- ◆ Insert an indwelling urinary catheter. Administer dextrose 5% in water or lactated Ringer solution, and antibiotics, as ordered. Carefully monitor nitroprusside I.V., if ordered; use a separate I.V. line for infusion. Adjust the dose by slowly increasing the infusion rate. Meanwhile, check blood pressure every 5 minutes until it stabilizes. With suspected bleeding from aneurysm, give whole blood transfusion.
- ◆ Explain diagnostic tests. If surgery is scheduled, explain the procedure and expected postoperative care (I.V. lines, ET and drainage tubes,

cardiac monitoring, and ventilation).

After repair of thoracic aneurysm:

- ◆ Assess LOC. Monitor vital signs; PAP, PAWP, and CVP; pulse rate; urine output; and pain.
- ◆ Check respiratory function. Carefully observe and record type and amount of chest tube drainage, and frequently assess heart and breath sounds.
- ◆ Monitor I.V. therapy.
- ◆ Give medications as appropriate.
- ◆ Watch for signs of infection, especially fever, and excessive wound drainage.
- ◆ Assist with range-of-motion exercises of legs to prevent thromboembolic phenomenon due to venostasis during prolonged bed rest.
- ◆ After stabilization of vital signs and respiration, encourage and assist the patient in turning, coughing, and deep breathing. If necessary, provide intermittent positive-pressure breathing to promote lung expansion. Help the patient walk as soon as he or she is able.
- ◆ Before discharge, ensure compliance with antihypertensive therapy by explaining the need for such drugs and the expected adverse effects. Teach the patient how to monitor blood pressure. Refer to community agencies for continued support and assistance, as needed.
- ◆ Throughout hospitalization, offer the patient and family psychological support. Answer all of their questions honestly and provide reassurance.

## **ABDOMINAL ANEURYSM**

### **Causes and Incidence**

Abdominal aneurysm, an abnormal dilation in the arterial wall, generally occurs in the aorta between the renal arteries and the iliac branches. Rupture—in which the aneurysm breaks open, resulting in profuse bleeding—is a common complication that occurs in larger aneurysms. Dissection occurs when the artery's lining tears, and blood leaks into the walls.

Abdominal aortic aneurysms (AAAs) result from arteriosclerosis, hypertension, congenital weakening, cystic medial necrosis, trauma, syphilis, and other infections. In children, this disorder can result from blunt abdominal injury or Marfan syndrome.

This disorder is four times more common in men than in women and is most prevalent in whites 40 to 70 years old. It is especially prevalent in male smokers who are 65 and older. Less than 50% of people with a ruptured AAA survive.

## **Pathophysiology**

These aneurysms develop slowly. First, a focal weakness in the muscular layer of the aorta (tunica media), due to degenerative changes, allows the inner layer (tunica intima) and outer layer (tunica adventitia) to stretch outward. Blood pressure within the aorta progressively weakens the vessel walls and enlarges the aneurysm.

## **Complications**

- ◆ Rupture
- ◆ Hemorrhage
- ◆ Shock

## **Signs and Symptoms**

Although abdominal aneurysms usually don't produce symptoms, most are evident (unless the patient is obese) as a pulsating mass in the periumbilical area, accompanied by a systolic bruit over the aorta. Some tenderness may be present on deep palpation. A large aneurysm may produce symptoms that mimic renal calculi, lumbar disk disease, and duodenal compression. Abdominal aneurysms rarely cause diminished peripheral pulses or claudication, unless embolization occurs.

Lumbar pain that radiates to the flank and groin from pressure on lumbar nerves may signify enlargement and imminent rupture. A rare but recognized symptom is unrelenting testicular pain with no other cause. If the aneurysm ruptures into the peritoneal cavity, it causes severe, persistent abdominal and back pain, mimicking renal or ureteral colic. Signs of hemorrhage—such as weakness, sweating, tachycardia, and hypotension—may be subtle because rupture into the retroperitoneal space produces a tamponade effect that prevents continued hemorrhage. Patients with such rupture may remain stable for hours before shock and death occur, although 20% die immediately.

## **Diagnosis**

Because abdominal aneurysms seldom produce symptoms, they're commonly detected accidentally as the result of an X-ray or a routine physical examination.



**CONFIRMING DIAGNOSIS** Several tests can confirm a suspected abdominal aneurysm. Serial ultrasound (sonography) can accurately determine the aneurysm's size, shape, and location. Anteroposterior and lateral X-rays of the abdomen can detect aortic calcification, which outlines the mass, at least 75% of the time. Aortography shows the condition of vessels proximal and distal to the aneurysm and the aneurysm's extent but may underestimate aneurysm diameter because it visualizes only the flow channel and not the surrounding clot. Computed tomography scan is used to diagnose and size the aneurysm. MRI can be used as an alternative to aortography.

## Treatment

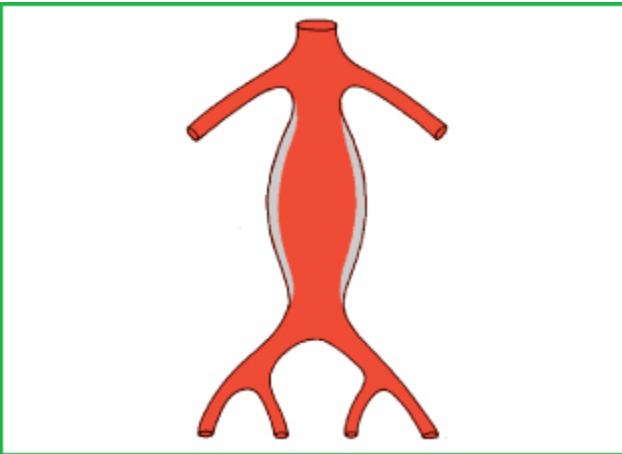
Usually, abdominal aneurysm requires resection of the aneurysm and replacement of the damaged aortic section with a Dacron graft. (See *Abdominal aneurysms: Before and after surgery*, page 75. Also see *Endovascular grafting for repair of an AAA*, page 76.) If the aneurysm is small and asymptomatic, surgery may be delayed, and the aneurysm may be followed and allowed to expand to a certain size because of possible surgical complications; however, small aneurysms may also rupture. Because of this risk, surgical repair or replacement is recommended for symptomatic patients or for patients with aneurysms greater than 5 cm in diameter.

## Abdominal Aneurysms: Before and After Surgery

During surgery, a prosthetic graft replaces or encloses the weakened area.

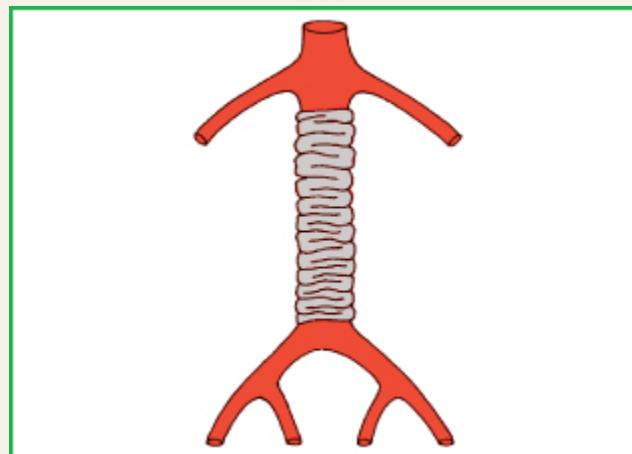
### Before Surgery

Aneurysm below renal arteries and above bifurcation



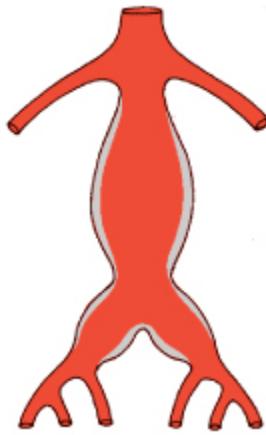
### **Before Surgery**

Aneurysm below renal arteries involving the iliac branches



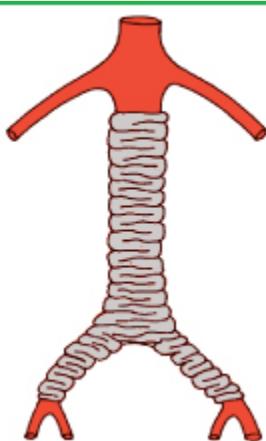
### **Before Surgery**

Small aneurysm in a patient with poor distal runoff (poor risk)



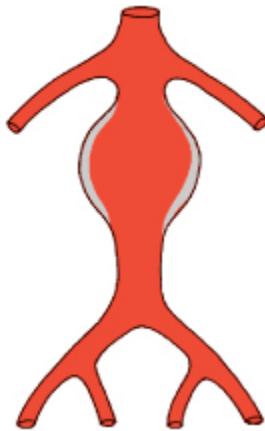
### **After Surgery**

The prosthesis extends distal to the renal arteries to above the aortic bifurcation.



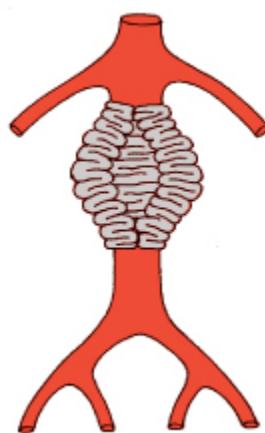
### **After Surgery**

The prosthesis extends to the common femoral arteries.



### After Surgery

The external prosthesis encircles the aneurysm and is held in place with sutures.



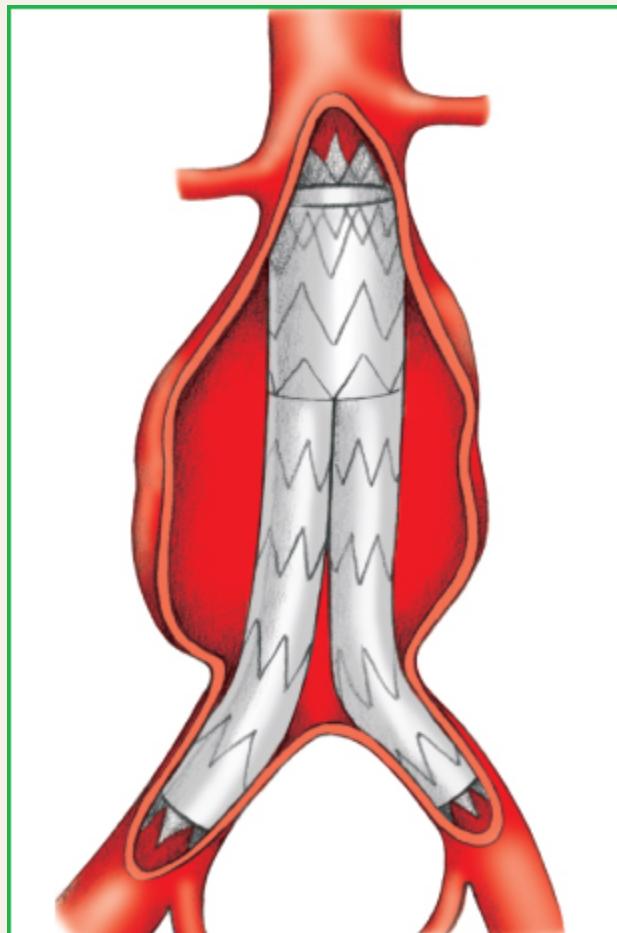
### Endovascular Grafting For Repair of An AAA

Endovascular grafting is a minimally invasive procedure for the repair of an abdominal aortic aneurysm (AAA). This procedure reinforces the walls of the aorta to prevent rupture and prevent expansion of the aneurysm.

Endovascular grafting is performed with fluoroscopic guidance: Using a guide wire, a delivery catheter with an attached compressed

graft is inserted through a small incision into the femoral or iliac artery. The delivery catheter is advanced into the aorta, where it's positioned across the aneurysm. A balloon on the catheter expands the graft and affixes it to the vessel wall.

The procedure generally takes 2 to 3 hours to perform. Patients are instructed to walk the first day after surgery and are generally discharged from the facility in 1 to 3 days.



Stenting is also a treatment option. It can be performed without an abdominal incision by introducing the catheters through arteries in the groin. However, not all patients with AAAs are candidates for this treatment.

Regular physical examination and ultrasound checks are necessary to detect enlargement, which may forewarn rupture. Large aneurysms or those

that produce symptoms pose a significant risk of rupture and necessitate immediate repair. In patients with poor distal runoff, external grafting may be done.

Risk factor modification is fundamental in the medical management of abdominal aneurysm, including control of hypocholesterolemia and hypertension. BBs are commonly prescribed to reduce the risk of aneurysm expansion and rupture.

## Special Considerations

Abdominal aneurysm requires meticulous preoperative and postoperative care, psychological support, and comprehensive patient teaching. Following diagnosis, if rupture isn't imminent, elective surgery allows time for additional preoperative tests to evaluate the patient's clinical status.

- ◆ Monitor vital signs, and type and crossmatch blood.
- ◆ Use only gentle abdominal palpation.
- ◆ As ordered, obtain renal function tests (BUN, creatinine, and electrolyte levels), blood samples (complete blood count with differential), electrocardiogram and cardiac evaluation, baseline pulmonary function tests, and ABG analysis.
- ◆ Be alert for signs of rupture, which may be immediately fatal. Watch closely for signs of acute blood loss (decreasing blood pressure; increasing pulse and respiratory rate; cool, clammy skin; restlessness; and decreased sensorium).
- ◆ If rupture does occur, the first priority is to get the patient to surgery immediately. Surgery allows direct compression of the aorta to control hemorrhage. Large amounts of blood may be needed during the resuscitative period to replace blood loss. In such a patient, renal failure caused by ischemia is a major postoperative complication, possibly requiring hemodialysis.
- ◆ Before elective surgery, weigh the patient, insert an indwelling urinary catheter and an I.V. line, and assist with insertion of an arterial line and pulmonary artery catheter to monitor fluid and hemodynamic balance. Give prophylactic antibiotics as ordered.
- ◆ Explain the surgical procedure and the expected postoperative care in the ICU for patients undergoing complex abdominal surgery (I.V. lines, ET and NG intubation, and mechanical ventilation).

- ◆ After surgery, in the ICU, closely monitor vital signs, intake and hourly output, neurologic status (LOC, pupil size, and sensation in arms and legs), and ABG values. Assess the depth, rate, and character of respirations and breath sounds at least every hour.
- ◆ Watch for signs of bleeding (increased pulse and respiratory rates and hypotension) and back pain, which may indicate that the graft is tearing. Check abdominal dressings for excessive bleeding or drainage. Be alert for temperature elevations and other signs of infection. After NG intubation for intestinal decompression, irrigate the tube frequently to ensure patency. Record the amount and type of drainage.
- ◆ Suction the ET tube often. If the patient can breathe unassisted and has good breath sounds and adequate ABG values, tidal volume, and vital capacity 24 hours after surgery, it is likely he or she will be extubated and will require oxygen by mask.
- ◆ Weigh the patient daily to evaluate fluid balance.
- ◆ Help the patient walk as soon as able (generally the second day after surgery).
- ◆ Provide psychological support for the patient and family. Help ease their fears about the ICU, the threat of impending rupture, and surgery by providing appropriate explanations and answering all questions.

## FEMORAL AND POPLITEAL ANEURYSMS

### Causes and Incidence

Femoral and popliteal aneurysms (sometimes called *peripheral arterial aneurysms*) are the end result of progressive atherosclerotic changes occurring in the walls (medial layer) of these major peripheral arteries. These aneurysmal formations may be *fusiform* (spindle-shaped) or *saccular* (pouchlike); the fusiform type is three times more common. They may be singular or multiple segmental lesions, often affecting both legs, and may accompany other arterial aneurysms located in the abdominal aorta or iliac arteries.

Femoral and popliteal aneurysms are usually secondary to atherosclerosis. Rarely, they result from congenital weakness in the arterial wall. They may also result from trauma (blunt or penetrating), bacterial infection, or peripheral vascular reconstructive surgery (which causes

“suture line” aneurysms, or false aneurysms, in which a blood clot forms a second lumen).

This condition occurs most frequently in men older than 50. The clinical course is usually progressive, eventually ending in thrombosis, embolization, and gangrene. Elective surgery before complications arise greatly improves the prognosis.

## **Pathophysiology**

As with other aneurysms, a weakening of the vessel wall caused by inflammation, proteolysis, and changes in the matrix result in the outpouching seen in this condition.

## **Complications**

- ◆ Amputation of thrombosis
- ◆ Emboli
- ◆ Gangrene

## **Signs and Symptoms**

Popliteal aneurysms may cause pain in the popliteal space when they’re large enough to compress the medial popliteal nerve and edema and venous distention if the vein is compressed. Femoral and popliteal aneurysms can produce symptoms of severe ischemia in the leg or foot due to acute thrombosis within the aneurysmal sac, embolization of mural thrombus fragments and, rarely, rupture. Symptoms of acute aneurysmal thrombosis include severe pain, loss of pulse and color, coldness in the affected leg or foot, and gangrene. Distal petechial hemorrhages may develop from aneurysmal emboli.

## **Diagnosis**

Diagnosis is usually confirmed by bilateral palpation that reveals a pulsating mass above or below the inguinal ligament in femoral aneurysm. When thrombosis has occurred, palpation detects a firm, nonpulsating mass. Arteriography or ultrasound may be indicated in doubtful situations. Arteriography may also detect associated aneurysms, especially those in the abdominal aorta and the iliac arteries. Ultrasound may be helpful in determining the size of the popliteal or femoral artery.

## Treatment

Femoral and popliteal aneurysms require surgical bypass and reconstruction of the artery, usually with an autogenous saphenous vein graft replacement. Arterial occlusion that causes severe ischemia and gangrene may require leg amputation.

## Special Considerations

Before corrective surgery:

- ◆ Assess and record circulatory status, noting the location and quality of peripheral pulses in the affected arm or leg.
- ◆ Administer prophylactic antibiotics or anticoagulants, as ordered.
- ◆ Discuss postoperative procedures and review the explanation of the surgery.

After arterial surgery:

- ◆ Monitor carefully for early signs of thrombosis or graft occlusion (loss of pulse, decreased skin temperature and sensation, and severe pain) and infection (fever).
- ◆ Palpate distal pulses at least every hour for the first 24 hours and then as frequently as ordered. Correlate these findings with preoperative circulatory assessment. Mark the sites on the patient's skin where pulses are palpable to facilitate repeated checks.
- ◆ Help the patient walk soon after surgery to prevent venostasis and possible thrombus formation.

To prepare the patient for discharge:

- ◆ Tell the patient to immediately report any recurrence of symptoms because the saphenous vein graft replacement can fail or another aneurysm may develop.
- ◆ Explain to the patient with popliteal artery resection that swelling may persist for some time. If antiembolism stockings are ordered, make sure they fit properly and teach the patient how to apply them. Warn against wearing constrictive apparel.
- ◆ If the patient is receiving anticoagulants, suggest measures to prevent bleeding, such as using an electric razor. Tell the patient to report any signs of bleeding (bleeding gums, tarry stools, and easy bruising) immediately. Explain the importance of follow-up blood studies to

monitor anticoagulant therapy. Warn the patient to avoid trauma, tobacco, and aspirin.

## THROMBOPHLEBITIS

### Causes and Incidence

An acute condition characterized by inflammation and thrombus formation, thrombophlebitis may occur in deep (intermuscular or intramuscular) or superficial (subcutaneous) veins. DVT or thrombophlebitis affects small veins, such as the soleal venous sinuses, or large veins, such as the vena cava and the femoral, iliac, and subclavian veins, causing venous insufficiency. (See *Chronic venous insufficiency*, page 78.) This disorder is typically progressive, leading to pulmonary embolism, a potentially lethal complication. Superficial thrombophlebitis is usually self-limiting and seldom leads to pulmonary embolism. Thrombophlebitis often begins with localized inflammation alone (phlebitis), but such inflammation rapidly provokes thrombus formation. Rarely, venous thrombosis develops without associated inflammation of the vein (phlebothrombosis).

### Chronic Venous Insufficiency

Chronic venous insufficiency results from the valvular destruction of deep vein thrombophlebitis, usually in the iliac and femoral veins, and occasionally the saphenous veins. It's often accompanied by incompetence of the communicating veins at the ankle, causing increased venous pressure and fluid migration into the interstitial tissue. Clinical effects include chronic swelling of the affected leg from edema, leading to tissue fibrosis, and induration; skin discoloration from extravasation of blood in subcutaneous tissue; and stasis ulcers around the ankle.

Treatment of small ulcers includes bed rest, elevation of the legs, warm soaks, and antimicrobial therapy for infection. Treatment to counteract increased venous pressure, the result of reflux from the deep venous system to surface veins, may include compression dressings, such as a sponge rubber pressure dressing or a zinc gelatin boot (Unna

boot). This therapy begins after massive swelling subsides with leg elevation and bed rest.

Large stasis ulcers unresponsive to conservative treatment may require excision and skin grafting. Patient care includes daily inspection to assess healing. Other care measures are the same as for varicose veins.

DVT may be idiopathic, but it usually results from endothelial damage, accelerated blood clotting, and reduced blood flow, known as the *Virchow triad*. Predisposing factors are prolonged bed rest, trauma, surgery, childbirth, and use of hormonal contraceptives such as estrogens. It occurs in about 80 of every 100,000 people; 1 of every 20 persons is affected at some point during their lifetime. Males are at slightly greater risk than females. People older than 40 are also at increased risk.

Causes of superficial thrombophlebitis include trauma, infection, I.V. drug abuse, and chemical irritation due to extensive use of the I.V. route for medications and diagnostic tests.

## Pathophysiology

A thrombus occurs when an alteration in the epithelial lining causes platelet aggregation and consequent fibrin entrapment of red and white blood cells and additional platelets. Thrombus formation is more rapid in areas where blood flow is slower, due to greater contact between platelet and thrombin accumulation. The rapidly expanding thrombus initiates a chemical inflammatory process in the vessel epithelium, which leads to fibrosis. The enlarging clot may occlude the vessel lumen partially or totally, or it may detach and embolize to lodge elsewhere in the systemic circulation.

## Complications

- ◆ Pulmonary embolism
- ◆ Chronic venous insufficiency

## Signs and Symptoms

In both types of thrombophlebitis, clinical features vary with the site and length of the affected vein. Although DVT may occur asymptotically, it may also produce severe pain, fever, chills, malaise and, possibly, swelling

and cyanosis of the affected arm or leg. Superficial thrombophlebitis produces visible and palpable signs, such as heat, pain, swelling, rubor, tenderness, and induration along the length of the affected vein. Varicose veins may also be present. (See *Varicose veins*, page 79.) Extensive vein involvement may cause lymphadenitis.

## Varicose Veins

Varicose veins are dilated, tortuous veins, usually affecting the subcutaneous leg veins—the saphenous veins and their branches. They can result from congenital weakness of the valves or venous wall, diseases of the venous system such as deep vein thrombophlebitis, conditions that produce prolonged venostasis such as pregnancy, or occupations that necessitate standing for an extended period.

Varicose veins may be asymptomatic or may produce mild to severe leg symptoms, including a feeling of heaviness; cramps at night; diffuse, dull aching after prolonged standing or walking; aching during menses; fatigability; palpable nodules, and, with deep vein incompetency, orthostatic edema and stasis pigmentation of the calves and ankles.

## Treatment

In mild-to-moderate varicose veins, antiembolism stockings or elastic bandages counteract pedal and ankle swelling by supporting the veins and improving circulation. An exercise program such as walking promotes muscular contraction and forces blood through the veins, thereby minimizing venous pooling. Severe varicose veins may necessitate stripping and ligation or, as an alternative to surgery, injection of a sclerosing agent into small affected vein segments.

To promote comfort and minimize worsening of varicosities:

- ◆ Discourage the patient from wearing constrictive clothing.
- ◆ Advise the patient to elevate the legs above heart level whenever possible and to avoid prolonged standing or sitting.

After stripping and ligation or after injection of a sclerosing agent:

- ◆ To relieve pain, administer analgesics as ordered.
- ◆ Frequently check circulation in toes (color and temperature) and observe elastic bandages for bleeding. When ordered, rewrap

bandages at least once a shift, wrapping from toe to thigh, with the leg elevated.

- ◆ Watch for signs of complications, such as sensory loss in the leg (which could indicate saphenous nerve damage), calf pain (thrombophlebitis), and fever (infection).

## Diagnosis

Findings are usually nonspecific and are not reliable for making the diagnosis of DVT. Essential laboratory tests include:

- ◆ Duplex Doppler is most commonly performed; this makes it possible to noninvasively examine the major veins (but not calf veins).



**CONFIRMING DIAGNOSIS** *Compression ultrasonography with Doppler is the diagnostic test of choice in the evaluation of DVT.*

Diagnosis must also rule out PAD, lymphangitis, cellulitis, and myositis.

Diagnosis of superficial thrombophlebitis is based on physical examination (redness and warmth over the affected area, palpable vein, and pain during palpation or compression).

## Treatment

The goals of treatment are to control thrombus development, prevent complications, relieve pain, and prevent recurrence of the disorder. Symptomatic measures include bed rest, with elevation of the affected arm or leg; warm, moist soaks to the affected area; and analgesics. After the acute episode of DVT subsides, the patient may resume activity while wearing antiembolism stockings that were applied before getting out of bed.

Treatment also includes anticoagulants to prolong clotting time. While warfarin is still an option for the treatment of DVT, new medications are now available that are safer, as effective, and do not require monitoring of blood levels through frequent lab draws. These medications are called direct factor Xa inhibitors and include rivaroxaban and apixaban. If the patient is not a candidate for these novel medications, low-molecular-weight (LMW) heparin has been shown to be effective in treating DVT or can be used as bridge therapy until a therapeutic level of warfarin is achieved. Although LMW heparin is more expensive, it doesn't require monitoring for its

anticoagulant effect, either. Full anticoagulant doses must be discontinued during any operative period because of the risk of hemorrhage. After some types of surgery, especially major abdominal or pelvic operations, prophylactic doses of anticoagulants may reduce the risk of DVT and pulmonary embolism. For lysis of acute, extensive DVT, treatment may include thrombolysis with or without thrombectomy. Rarely, DVT may cause complete venous occlusion, which necessitates venous interruption through simple ligation to vein plication, or clipping. Embolectomy and insertion of a vena caval umbrella or filter may also be done.

Therapy for severe superficial thrombophlebitis may include an anti-inflammatory drug such as indomethacin, antiembolism stockings, warm soaks, and elevation of the leg.

## **Special Considerations**

Patient teaching, identification of high-risk patients, and measures to prevent venostasis can prevent DVT; close monitoring of anticoagulant therapy can prevent serious complications such as internal hemorrhage.

- ◆ Enforce bed rest as ordered, and elevate the patient's affected arm or leg. If you plan to use pillows for elevating the leg, place them so they support the entire length of the affected extremity to prevent possible compression of the popliteal space.
- ◆ Apply warm soaks to increase circulation to the affected area and to relieve pain and inflammation. Give analgesics to relieve pain, as ordered.
- ◆ Measure and record the affected arm or leg's circumference daily, and compare this measurement to the other arm or leg. To ensure accuracy and consistency of serial measurements, mark the skin over the area and measure at the same spot daily.
- ◆ Administer heparin I.V., as ordered, with an infusion monitor or pump to control the flow rate if necessary. Remember that this medication is not being used as often with the growing popularity of direct factor Xa inhibitors.
- ◆ Measure partial thromboplastin time regularly for the patient on heparin therapy; prothrombin time and international normalized ratio (INR) for the patient on warfarin (therapeutic anticoagulation values are 1½ to 2 times control values for prothrombin time and an INR of 2 to 3). Watch for signs and symptoms of bleeding, such as dark, tarry stools; coffee-

ground vomitus; and ecchymosis. Encourage the patient to use an electric razor and to avoid medications that contain aspirin.

- ◆ Be alert for signs of pulmonary emboli (crackles, dyspnea, hemoptysis, sudden changes in mental status, restlessness, and hypotension).

To prepare the patient with thrombophlebitis for discharge:

- ◆ Emphasize the importance of follow-up blood studies to monitor anticoagulant therapy, if necessary.
- ◆ If the patient is being discharged on heparin therapy, teach the patient or family how to give subcutaneous injections. If the patient requires further assistance, arrange for a home health nurse.
- ◆ Tell the patient to avoid prolonged sitting or standing to help prevent recurrence.
- ◆ Teach the patient how to properly apply and use antiembolism stockings. Tell the patient to report any complications such as cold, blue toes. (See *Preventing thrombophlebitis*.)



## PREVENTION PREVENTING THROMBOPHLEBITIS

To prevent thrombophlebitis in a high-risk patient, perform range-of-motion exercise while the patient is on bed rest, use intermittent pneumatic calf massage during lengthy surgical or diagnostic procedures, apply antiembolism stockings postoperatively, and encourage early ambulation.

After some types of surgery, especially major abdominal or pelvic operations, prophylactic doses of anticoagulants may reduce the risk of deep vein thrombosis and pulmonary embolism.

## RAYNAUD DISEASE

### Causes and Incidence

Raynaud disease is one of several primary arteriospastic disorders characterized by episodic vasospasm in the small peripheral arteries and arterioles, precipitated by exposure to cold or stress. This condition occurs

bilaterally and usually affects the hands or, less often, the feet. Raynaud disease is most prevalent in females, particularly those between puberty and 40 years old. It's a benign condition, requiring no specific treatment and causing no serious sequelae.

Although the cause is unknown, several theories account for the reduced digital blood flow: intrinsic vascular wall hyperactivity to cold, increased vasomotor tone due to sympathetic stimulation, and antigen–antibody immune response (the most likely theory because abnormal immunologic test results accompany Raynaud phenomenon). Risk factors include associated diseases (Buerger disease, atherosclerosis, rheumatoid arthritis, scleroderma, and SLE) and smoking.

This disorder affects females more often than males.

Raynaud phenomenon, however, a condition commonly associated with several connective tissue disorders—such as scleroderma, SLE, or polymyositis—has a progressive course, leading to ischemia, gangrene, and amputation. Distinguishing between the two disorders is difficult because some patients who experience mild symptoms of Raynaud disease for several years may later develop overt connective tissue disease—especially scleroderma.

## Pathophysiology

## Complications

- ◆ Ischemia
- ◆ Gangrene
- ◆ Amputation

## Signs and Symptoms

After exposure to cold or stress, the skin on the fingers typically blanches and then becomes cyanotic before changing to red and before changing from cold to normal temperature. Numbness and tingling may also occur. These symptoms are relieved by warmth. In long-standing disease, trophic changes, such as sclerodactyly, ulcerations, or chronic paronychia, may result. Although it's extremely uncommon, minimal cutaneous gangrene necessitates amputation of one or more phalanges.

## Diagnosis

Clinical criteria that establish Raynaud disease include skin color changes induced by cold or stress; bilateral involvement; absence of gangrene or, if present, minimal cutaneous gangrene; normal arterial pulses; and patient history of clinical symptoms of longer than 2 years' duration. Diagnosis must also rule out secondary disease processes, such as chronic arterial occlusive or connective tissue disease.

## Treatment

Initially, treatment consists of avoidance of cold, mechanical, or chemical injury; cessation of smoking; and reassurance that symptoms are benign. Because adverse drug effects, especially from vasodilators, may be more bothersome than the disease itself, drug therapy is reserved for unusually severe symptoms. Such therapy may include low doses of nifedipine. Sympathectomy may be helpful when conservative modalities fail to prevent ischemic ulcers and becomes necessary in less than 25% of patients.

## Special Considerations

- ◆ Warn the patient against exposure to the cold. Tell the patient to wear mittens or gloves in cold weather or when handling cold items or defrosting the freezer.
- ◆ Advise the patient to avoid stressful situations and to stop smoking.
- ◆ Instruct the patient to inspect the skin frequently and to seek immediate care for signs of skin breakdown or infection.
- ◆ Teach the patient about drugs, their use, and their adverse effects.
- ◆ Provide psychological support and reassurance to allay the patient's fear of amputation and disfigurement.

## BUERGER DISEASE

### Causes and Incidence

Buerger disease (sometimes called *thromboangiitis obliterans*)—an inflammatory, nonatheromatous occlusive condition—causes segmental lesions and subsequent thrombus formation in the small and medium arteries (and sometimes the veins), resulting in decreased blood flow to the feet and legs. This disorder may produce ulceration and, eventually, gangrene.

Buerger disease is caused by vasculitis, an inflammation of blood vessels, primarily of the hands and feet. The vessels become constricted or totally blocked, reducing blood flow to the tissues and resulting in pain and, eventually, damage.

This disorder occurs in 12 to 20 of every 100,000 people. Incidence is highest among males 20 to 40 years old who have a history of smoking or chewing tobacco. It may be associated with a history of Raynaud disease and may occur in people with autoimmune disease.

## **Pathophysiology**

The pathophysiology of Buerger disease is not well understood. In the acute phase, occlusive thrombi develop in arteries and veins of distal extremities. Next, thrombi start to organize into larger vessels. Over time, inflammation resides but fibrosis and organized thrombi remain.

There is also evidence that dysfunction of the endothelial layer of the vessels occurs, as well as the possibility of issues with prothrombin.

## **Complications**

- ◆ Gangrene
- ◆ Muscle atrophy
- ◆ Ulceration

## **Signs and Symptoms**

Buerger disease typically produces intermittent claudication of the instep, which is aggravated by exercise and relieved by rest. During exposure to low temperature, the feet initially become cold, cyanotic, and numb; later, they redden, become hot, and tingle. Occasionally, Buerger disease also affects the hands, possibly resulting in painful fingertip ulcerations. Associated signs and symptoms may include impaired peripheral pulses, migratory superficial thrombophlebitis and, in later stages, ulceration, muscle atrophy, and gangrene.

## **Diagnosis**

Patient history and physical examination strongly suggest Buerger disease. Supportive diagnostic tests include:

- ◆ Doppler ultrasonography to show diminished circulation in the peripheral vessels
- ◆ angiography or arteriography to locate lesions and rule out atherosclerosis.

## Treatment

The primary goals of treatment are to relieve symptoms and prevent complications. Such therapy may include an exercise program that uses gravity to fill and drain the blood vessels or, in severe disease, a lumbar sympathectomy to increase blood supply to the skin. Aspirin and vasodilators may also be used. Amputation may be necessary for nonhealing ulcers, intractable pain, or gangrene.

## Special Considerations

- ◆ Strongly urge the patient to stop smoking to enhance the treatment's effectiveness. Symptoms may disappear if the patient stops tobacco use. If necessary, refer the patient to a self-help group to stop smoking.
- ◆ Warn the patient to avoid precipitating factors, such as emotional stress, exposure to extreme temperatures, and trauma.
- ◆ Teach the patient proper foot care, especially the importance of wearing well-fitting shoes and cotton or wool socks. Show the patient how to inspect feet daily for cuts, abrasions, and signs of skin breakdown, such as redness and soreness. Remind the patient to seek medical attention at once after any trauma.
- ◆ If the patient has ulcers and gangrene, enforce bed rest and use a padded footboard or bed cradle to prevent pressure from bed linens. Protect the feet with soft padding. Wash them gently with a mild soap and tepid water, rinse thoroughly, and pat dry with a soft towel.
- ◆ Provide emotional support. If necessary, refer the patient for psychological counseling to help the patient cope with restrictions imposed by this chronic disease. If the patient has undergone amputation, assess rehabilitative needs, especially regarding changes in body image. Refer the patient to physical therapists, occupational therapists, and social service agencies, as needed.

## PERIPHERAL ARTERY DISEASE

## Causes and Incidence

PAD, referred to as *arterial occlusive disease*, is the obstruction or narrowing of the lumen of the aorta and its major branches, causing an interruption of blood flow, usually to the legs and feet. PAD may affect the carotid, vertebral, innominate, subclavian, mesenteric, and celiac arteries. Occlusions may be acute or chronic and commonly cause severe ischemia, skin ulceration, and gangrene.

The prognosis depends on the occlusion's location, the development of collateral circulation to counteract reduced blood flow, and, in acute disease, the time elapsed between occlusion and its removal.

Predisposing factors include smoking; aging; such conditions as hypertension, hyperlipidemia, and diabetes; and a family history of vascular disorders, MI, or stroke. PAD has no racial predilection. Men older than 50 are at increased risk for intermittent claudication, a common sign of PAD.

## Pathophysiology

PAD is a common complication of atherosclerosis. The occlusive mechanism may be endogenous, due to emboli formation or thrombosis, or exogenous, due to trauma or fracture.

## Complications

- ◆ Severe ischemia
- ◆ Skin ulceration
- ◆ Gangrene
- ◆ Limb loss

## Signs and Symptoms

The signs and symptoms of PAD depend on the site of the occlusion. (See *Types of peripheral artery disease*, page 81.)

### Types of Peripheral Artery Disease

| <b>Site of occlusion</b>       | <b>Signs and symptoms</b> |
|--------------------------------|---------------------------|
| <b>Carotid arterial system</b> |                           |

| <b>Site of occlusion</b>   | <b>Signs and symptoms</b>  |
|--|--|
| Internal carotids  | <ul style="list-style-type: none"> <li>◆ Absent or decreased pulsation with an auscultatory bruit over the affected vessels</li> </ul>   |
| External carotids  | <ul style="list-style-type: none"> <li>◆ Neurologic dysfunction: transient ischemic attacks (TIAs) due to reduced cerebral circulation producing unilateral sensory or motor dysfunction (transient monocular blindness, hemiparesis), possible aphasia or dysarthria, confusion, decreased mentation, and headache (These are recurrent features that usually last 5 to 10 minutes but may persist up to 24 hours and may herald a stroke)</li> </ul> |
| <b>Vertebrobasilar system</b>  |  |
| Vertebral arteries   | <ul style="list-style-type: none"> <li>◆ Neurologic dysfunction: TIAs of the brainstem and cerebellum producing binocular vision disturbances, vertigo, dysarthria, and “drop attacks” (falling down without loss of consciousness); less common than carotid TIA</li> </ul>   |
| Basilar arteries   |  |
| <b>Innominates</b>   |  |
| Brachiocephalic artery   | <ul style="list-style-type: none"> <li>◆ Indications of ischemia (claudication) of the right arm</li> <li>◆ Neurologic dysfunction: signs and symptoms of vertebrobasilar occlusion</li> <li>◆ Possible bruit over the right side of the neck</li> </ul>   |
| <b>Subclavian artery</b>   |  |
|  | <ul style="list-style-type: none"> <li>◆ Clinical effects of vertebrobasilar occlusion and exercise-induced arm claudication</li> <li>◆ Subclavian steal syndrome (characterized by the backflow of blood from the brain through the vertebral artery on the same side as the occlusion, into the subclavian artery distal to the occlusion)</li> <li>◆ Possibly gangrene (usually limited to the digits)</li> </ul>                                   |
| <b>Mesenteric artery</b>   |  |
| Superior (most commonly affected)  | <ul style="list-style-type: none"> <li>◆ Bowel ischemia, infarct necrosis, and gangrene</li> <li>◆ Diarrhea</li> <li>◆ Leukocytosis</li> </ul>   |
| Celiac axis  | <ul style="list-style-type: none"> <li>◆ Nausea and vomiting</li> </ul>  |
| Inferior   | <ul style="list-style-type: none"> <li>◆ Shock due to massive intraluminal fluid and plasma loss</li> <li>◆ Sudden, acute abdominal pain</li> </ul>  |
| <b>Aortic bifurcation</b>  |  |
| (Saddle block occlusion, a medical emergency associated with cardiac embolization) | <ul style="list-style-type: none"> <li>◆ Sensory and motor deficits (muscle weakness, numbness, paresthesias, and paralysis) in both legs</li> <li>◆ Signs of ischemia (sudden pain and cold, pale legs with decreased or absent peripheral pulses) in both legs</li> </ul>  |
| <b>Iliac artery</b>  |  |
| (Leriche syndrome)   | <ul style="list-style-type: none"> <li>◆ Absent or reduced femoral or distal pulses</li> <li>◆ Impotence</li> <li>◆ Intermittent claudication of the lower back, buttocks, and thighs, relieved by rest</li> <li>◆ Possible bruit over femoral arteries</li> </ul>   |

| <b>Site of occlusion</b>  | <b>Signs and symptoms</b>   |
|---|---|
| <b>Femoral and popliteal artery</b><br><br>(Associated with aneurysm formation) | <ul style="list-style-type: none"> <li>◆ Gangrene</li> <li>◆ Intermittent claudication of the calves on exertion</li> <li>◆ Ischemic pain in feet</li> <li>◆ Leg pallor and coolness; blanching of the feet on elevation</li> <li>◆ No palpable pulses in the ankles and feet</li> <li>◆ Pretrophic pain (heralds necrosis and ulceration)</li> </ul> |

## Diagnosis

Diagnosis of PAD is usually indicated by patient history and physical examination.

Pertinent supportive diagnostic tests include the following:

- ◆ Arteriography demonstrates the type (thrombus or embolus), location, and degree of obstruction and the collateral circulation. It's particularly useful in chronic disease or for evaluating candidates for reconstructive surgery.
- ◆ Doppler ultrasonography and plethysmography are noninvasive tests that show decreased blood flow distal to the occlusion in acute disease.
- ◆ EEG and computed tomography scan may be necessary to rule out brain lesions.
- ◆ Ankle-brachial indices compare the systolic blood pressures between the upper and lower extremities and have been shown to be sensitive in detecting PAD.

## Treatment

Treatment depends on the obstruction's cause, location, and size. For mild chronic disease, supportive measures include elimination of smoking, hypertension control, and walking exercise. For carotid artery occlusion, antiplatelet therapy may begin with clopidogrel and aspirin. For intermittent claudication of chronic occlusive disease, cilostazol may improve blood flow through the capillaries, particularly for patients who are poor candidates for surgery. Statin medications are used frequently, as well, to lessen the progression of atherosclerosis.

Acute PAD usually requires surgery to restore circulation to the affected area, for example:

- ◆ Atherectomy—Excision of plaque using a drill or slicing mechanism.
- ◆ Balloon angioplasty—Compression of the obstruction using balloon inflation.
- ◆ Bypass graft—Blood flow is diverted through an anastomosed autogenous or Dacron graft past the thrombosed segment.
- ◆ Combined therapy—Concomitant use of any of the above treatments.
- ◆ Embolectomy—A balloon-tipped Fogarty catheter is used to remove thrombotic material from the artery. Embolectomy is used mainly for mesenteric, femoral, or popliteal artery occlusion.
- ◆ Laser angioplasty—Use of excision and hot-tipped lasers to vaporize the obstruction.
- ◆ Lumbar sympathectomy—An adjunct to surgery, depending on the sympathetic nervous system's condition.
- ◆ Patch grafting—Removal of the thrombosed arterial segment and replacement with an autogenous vein or Dacron graft.
- ◆ Stents—Insertion of a mesh of wires that stretch and mold to the arterial wall to prevent reocclusion. This follows laser angioplasty or atherectomy.
- ◆ Thromboendarterectomy—Opening of the occluded artery and direct removal of the obstructing thrombus and the medial layer of the arterial wall; usually performed after angiography and commonly used with autogenous vein or Dacron bypass surgery (femoral-popliteal or aortofemoral).
- ◆ Thrombolytic therapy—Lysis of any clot around or in the plaque by urokinase, streptokinase, or alteplase.

Amputation becomes necessary with failure of arterial reconstructive surgery or with the development of gangrene, persistent infection, or intractable pain.

Other therapy includes heparin to prevent emboli (for embolic occlusion) and bowel resection after restoration of blood flow (for mesenteric artery occlusion).

## Special Considerations

- ◆ Provide comprehensive patient teaching, including proper foot care. Explain diagnostic tests and procedures. Advise the patient to stop smoking and to follow the prescribed medical regimen.

Preoperatively, during an acute episode:

- ◆ Assess the patient's circulatory status by checking for the most distal pulses and by inspecting skin color and temperature.
- ◆ Provide pain relief as needed.
- ◆ Administer heparin by continuous I.V. drip as ordered. Use an infusion monitor or pump to ensure the proper flow rate.
- ◆ Wrap the patient's affected foot in soft cotton batting, and reposition it frequently to prevent pressure on any one area. Strictly avoid elevating or applying heat to the affected leg.
- ◆ Watch for signs of fluid and electrolyte imbalance, and monitor intake and output for signs of renal failure (urine output <30 mL/hour).
- ◆ If the patient has carotid, innominate, vertebral, or subclavian artery occlusion, monitor for signs of stroke, such as numbness in an arm or leg and intermittent blindness.

Postoperatively:

- ◆ Monitor the patient's vital signs. Continuously assess circulatory function by inspecting skin color and temperature and by checking for distal pulses. In charting, compare earlier assessments and observations. Watch closely for signs of hemorrhage (tachycardia and hypotension) and check dressings for excessive bleeding.
- ◆ In carotid, innominate, vertebral, or subclavian artery occlusion, assess neurologic status frequently for changes in LOC or muscle strength and pupil size.
- ◆ In mesenteric artery occlusion, connect the NG tube to low intermittent suction. Monitor intake and output (low urine output may indicate damage to renal arteries during surgery). Check bowel sounds for return of peristalsis. Increasing abdominal distention and tenderness may indicate extension of bowel ischemia with resulting gangrene, necessitating further excision, or it may indicate peritonitis.
- ◆ In saddle block occlusion, check distal pulses for adequate circulation. Watch for signs of renal failure and mesenteric artery occlusion (severe abdominal pain), and for cardiac arrhythmias, which may precipitate embolus formation.
- ◆ In iliac artery occlusion, monitor urine output for signs of renal failure from decreased perfusion to the kidneys as a result of surgery. Provide meticulous catheter care.

- ◆ In both femoral and popliteal artery occlusions, assist with early ambulation, but discourage prolonged sitting.
- ◆ After amputation, check the patient's stump carefully for drainage; record the color and amount of damage, and the time. Elevate the stump as ordered, and administer adequate analgesic medication. Because phantom limb pain is common, explain this phenomenon to the patient.
- ◆ When preparing the patient for discharge, instruct the patient to watch for signs of recurrence (pain, pallor, numbness, paralysis, and absence of pulse) that can result from graft occlusion or occlusion at another site. Warn the patient against wearing constrictive clothing.

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# 2

## Respiratory Disorders

### Introduction

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The respiratory system distributes air to the alveoli, where gas exchange—the addition of oxygen ( $O_2$ ) and the removal of carbon dioxide ( $CO_2$ ) from pulmonary capillary blood—takes place. Certain specialized structures within this system play a vital role in preparing air for use by the body. The nose, for example, contains vestibular hairs that filter the air and an extensive vascular network that warms it. The nose also contains a layer of goblet cells and a moist mucosal surface; water vapor enters the airstream from this mucosal surface to saturate inspired air as it's warmed in the upper airways. Ciliated mucosa in the posterior portion of the nose and nasopharynx as well as major portions of the tracheobronchial tree propel particles deposited by impaction or gravity to the oropharynx, where the particles are swallowed.

### EXTERNAL RESPIRATION

The external component of respiration—ventilation or breathing—delivers inspired air to the lower respiratory tract and alveoli. Contraction and relaxation of the respiratory muscles move air into and out of the lungs. Ventilation begins with the contraction of the inspiratory muscles: the diaphragm (the major muscle of respiration) descends, while external intercostal muscles move the rib cage upward and outward.

Air then enters the lungs in response to the pressure gradient between the atmosphere and the lungs. The lungs adhere to the chest wall and diaphragm

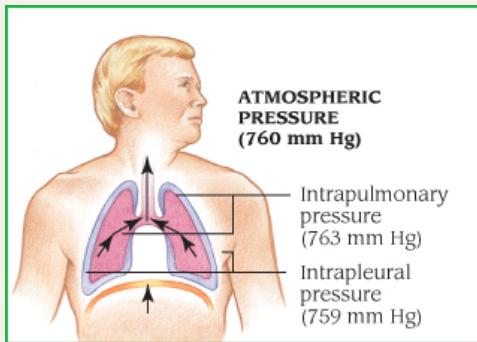
because of the vacuum created within the pleural space. As the thorax expands, negative pressure is created in the intrapleural space, causing the lungs to also expand and draw in the warmed, humidified air. The accessory muscles of inspiration, which include the scalene and sternocleidomastoid muscles, raise the clavicles, upper ribs, and sternum. The accessory muscles aren't used in normal inspiration but may be used in some pathologic conditions.

Normal expiration is passive; the inspiratory muscles cease to contract, the diaphragm rises, and the elastic recoil of the lungs causes the lungs to contract. These actions raise the pressure within the lungs above atmospheric pressure, moving air from the lungs to the atmosphere. Active expiration causes pleural pressure to become less negative. (See *Mechanics of ventilation*, page 87.)

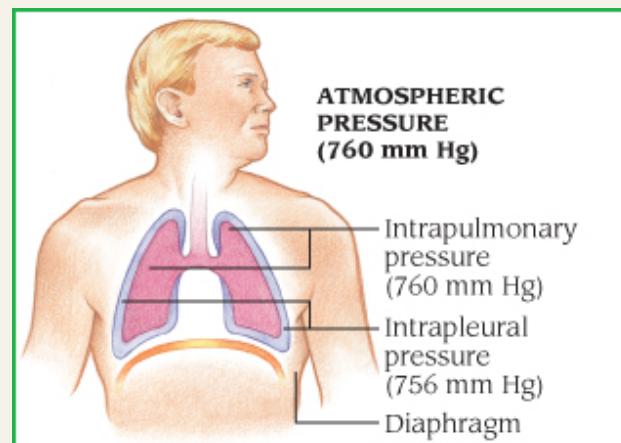
## Mechanics of Ventilation

Breathing results from differences between atmospheric and intrapulmonary pressures, as described below.

Before inspiration, intrapulmonary pressure equals atmospheric pressure (~760 mm Hg). Intrapleural pressure is 756 mm Hg.



The intrapulmonary atmospheric pressure gradient pulls air into the lungs until the two pressures are equal.

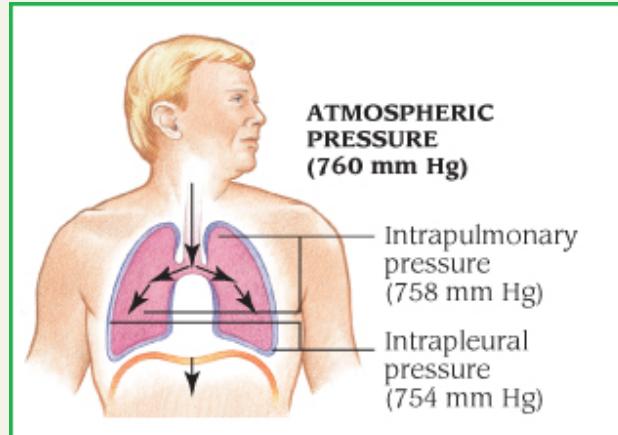
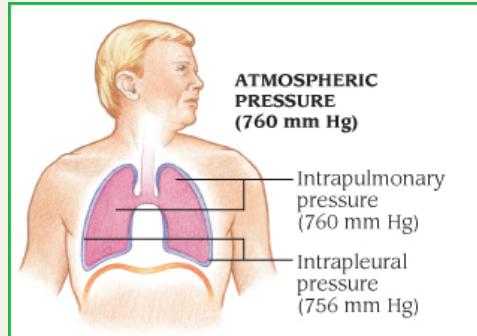


During inspiration, the diaphragm and external intercostal muscles contract,

During normal expiration, the diaphragm slowly relaxes and the lungs and thorax passively return to resting size and

enlarging the thorax vertically and horizontally. As the thorax expands, intrapleural pressure decreases and the lungs expand to fill the enlarging thoracic cavity.

During deep or forced expiration, contraction of internal intercostal and abdominal muscles reduces thoracic volume. Lung and thorax compression raises intrapulmonary pressure above atmospheric pressure.



An adult lung contains an estimated 300 million alveoli; each alveolus is supplied by many capillaries. To reach the capillary lumen, O<sub>2</sub> must cross the alveolocapillary membrane, which consists of an alveolar epithelial cell, a thin interstitial space, the capillary basement membrane, and the capillary endothelial cell membrane. The O<sub>2</sub> tension of air entering the respiratory tract is approximately 150 mm Hg. In the alveoli, inspired air mixes with CO<sub>2</sub> and water vapor, lowering the O<sub>2</sub> pressure to approximately 100 mm Hg. Because alveolar partial pressure of O<sub>2</sub> is higher than that present in mixed venous blood entering the pulmonary capillaries (~40 mm Hg), O<sub>2</sub> diffuses across the alveolocapillary membrane into the blood.

## O<sub>2</sub> AND CO<sub>2</sub> TRANSPORT AND INTERNAL RESPIRATION

Circulating blood delivers O<sub>2</sub> to the cells of the body for metabolism and transports metabolic wastes and CO<sub>2</sub> from the tissues back to the lungs. When oxygenated arterial blood reaches tissue capillaries, O<sub>2</sub> diffuses from the blood into the cells because of the O<sub>2</sub> tension gradient. The amount of O<sub>2</sub> available is determined by the concentration of hemoglobin (Hb; the principal

carrier of O<sub>2</sub>), the percentage of O<sub>2</sub> saturation of the Hb, regional blood flow, arterial O<sub>2</sub> content, and cardiac output.

Internal (cellular) respiration occurs as a part of cellular metabolism, which can take place with O<sub>2</sub> (aerobic) or without O<sub>2</sub> (anaerobic). The most efficient method for providing fuel (high-energy compounds such as adenosine triphosphate [ATP]) for cellular reactions is aerobic metabolism, which produces CO<sub>2</sub> and water in addition to ATP. Anaerobic metabolism is less efficient because a cell produces only a limited amount of ATP and yields lactic acid as well as CO<sub>2</sub> as a metabolic by-product.

Because circulation is continuous, CO<sub>2</sub> doesn't normally accumulate in tissues. CO<sub>2</sub> produced during cellular respiration diffuses from tissues into regional capillaries and is transported by systemic venous circulation. When CO<sub>2</sub> reaches the alveolar capillaries, it diffuses into the alveoli, where the partial pressure of CO<sub>2</sub> is lower; CO<sub>2</sub> is removed from the alveoli during exhalation.

## MECHANISMS OF CONTROL

The central nervous system's (CNS) control of respiration lies in the respiratory center, located in the lateral medulla oblongata of the brainstem. Impulses travel down the phrenic nerves to the diaphragm, and down the intercostal nerves to the intercostal muscles, where the impulses change the rate and depth of respiration. The inspiratory and expiratory centers, located in the posterior medulla, establish the involuntary rhythm of the breathing pattern.

Apneustic and pneumotaxic centers in the pons influence the pattern of breathing. Stimulation of the lower pontine apneustic center (e.g., by trauma, tumor, or stroke) produces forceful inspiratory gasps alternating with weak expiration. The apneustic center continually excites the medullary inspiratory center and thus facilitates inspiration. Signals from the pneumotaxic center as well as afferent impulses from the vagus nerve inhibit the apneustic center and "turn off" inspiration. The apneustic pattern doesn't occur if the vagus nerves are intact.

Partial pressure of arterial oxygen (Pao<sub>2</sub>), pH, and pH of cerebrospinal fluid (CSF) influence output from the respiratory center. When CO<sub>2</sub> enters the CSF, the pH of CSF falls, stimulating central chemoreceptors to increase ventilation.

The respiratory center also receives information from peripheral chemoreceptors in the carotid and aortic bodies. These chemoreceptors respond primarily to decreased  $\text{PaO}_2$  but also to decreased pH. The peripheral chemoreceptors have little control over respirations until the  $\text{PaO}_2$  is less than 60 mm Hg.

During exercise, stretch receptors in lung tissue and the diaphragm prevent overexpansion of the lungs. During swallowing, the cortex can interrupt automatic control of ventilation. During sleep, respiratory drive may fluctuate, producing hypoventilation and periods of apnea. External sensations, drugs, chronic hypercapnia, and changes in body temperature can also alter the respiratory pattern.

## DIAGNOSTIC TESTS

Diagnostic tests evaluate physiologic characteristics and pathologic states within the respiratory tract.

Noninvasive tests include:

- ◆ Chest X-ray shows such conditions as atelectasis, pleural effusion, infiltrates, pneumothorax, lesions, mediastinal shifts, pulmonary edema, and chronic obstructive pulmonary disease (COPD).
- ◆ Computed tomography (or CT) scan provides a three-dimensional picture that's 100 times more sensitive than a chest X-ray.
- ◆ Magnetic resonance imaging identifies obstructed arteries and tissue perfusion, but movement of the heart and lungs reduces the image's clarity.
- ◆ Sputum specimen analysis assesses sputum quantity, color, viscosity, and odor; microbiological stains and culture of sputum can identify infectious organisms; and cytologic preparations can detect respiratory tract neoplasms. Sensitivity tests determine antibiotic sensitivity and resistance.
- ◆ Pulmonary function tests (or PFTs) measure lung volume, flow rates, and compliance. Normal values, individualized by body stature, ethnicity, and age, are reported in percentage of the normal predicted value. Static measurements are volume measurements that include tidal volume, volume of air contained in a normal breath; functional residual capacity, volume of air remaining in the lungs after normal expiration; vital capacity, volume of air that can be exhaled after maximal inspiration; residual volume, air remaining in the lungs after maximal expiration; and total lung capacity (TLC), volume of air in the lungs after maximal inspiration. Dynamic measurements characterize the movement of air into and out of the lungs

and show changes in lung mechanics. They include measurement of forced expiratory volume in 1 second (FEV<sub>1</sub>), maximum volume of air that can be expired in 1 second from TLC; maximal voluntary ventilation, volume of air that can be expired in 1 minute with the patient's maximum voluntary effort; and forced vital capacity (FVC), maximal volume of air that the patient can exhale from TLC. (Peak flow rate, which can be obtained at the bedside, is also a dynamic measurement of pulmonary function.)

- ◆ Methacholine challenge is one method of assessing airway responsiveness and is used to determine a diagnosis of asthma.
- ◆ Exercise stress test evaluates the ability to transport O<sub>2</sub> and remove CO<sub>2</sub> with increasing metabolic demands.
- ◆ Polysomnography can diagnose sleep disorders.
- ◆ Lung scan (ventilation–perfusion or scintiphotography scan) demonstrates ventilation and perfusion patterns. It's used primarily to evaluate pulmonary embolus.
- ◆ Arterial blood gas (ABG) analysis assesses gas exchange. Decreased Pao<sub>2</sub> may indicate hypoventilation, ventilation–perfusion mismatch, or shunting of blood away from gas exchange sites. Increased partial pressure of arterial carbon dioxide (Paco<sub>2</sub>) reflects marked ventilation–perfusion mismatch or hypoventilation; decreased Paco<sub>2</sub> reflects increased alveolar ventilation. Changes in pH may reflect metabolic or respiratory dysfunction.
- ◆ Pulse oximetry is a noninvasive assessment of arterial oxygen saturation.
- ◆ Capnography may be used either transcutaneously or in ventilator circuit to determine Paco<sub>2</sub> trends.

Invasive tests include:

- ◆ Bronchoscopy permits direct visualization of the trachea and mainstem, lobar, segmental, and subsegmental bronchi. It may be used to localize the site of lung hemorrhage, visualize masses in these airways, and collect respiratory tract secretions. Brush biopsy may be used to obtain specimens from the lungs for microbiological stains, culture, and cytology. Lesion biopsies may be performed by using small forceps under direct visualization (when present in the proximal airways) or with the aid of fluoroscopy (when present distal to regions of direct visualization). Bronchoscopy can also be used to clear secretions and remove foreign bodies.

- ◆ Thoracentesis permits removal of pleural fluid for analysis.
- ◆ Pleural biopsy obtains pleural tissue for histologic examination and culture.
- ◆ Pulmonary artery angiography, the injection of dye into the pulmonary artery, can locate pulmonary embolism. This is considered the gold standard for diagnosing pulmonary emboli.
- ◆ Positron emission tomography scan uses a short-life radionuclide. Increased uptake of the substance is seen in malignant cells.

## **ASSESSMENT**

Assessment of the respiratory system begins with a thorough patient history. Ask the patient to describe his or her respiratory problem. How long has he or she had it? How long does each attack last? Does one attack differ from another? Does any activity in particular bring on an attack or make it worse? What relieves the symptoms? Always ask whether the patient was or is a smoker, what and how often he or she smoked or smokes, and how long he or she smoked or has been smoking. Record this information in *pack years*—the number of packs of cigarettes per day multiplied by the number of smoking years. Remember to ask about the patient's occupation, hobbies, and travel; some of these activities may involve exposure to toxic or allergenic substances.

If the patient has dyspnea, ask if it occurs during activity or at rest. What position is the patient in when dyspnea occurs? How far can he or she walk? How many flights of stairs can he or she climb? Has his or her exercise tolerance been decreasing? Can he or she relate dyspnea to allergies or environmental conditions? Does it occur only at night, during sleep? If the patient has a cough, ask about its severity, persistence, and duration; ask if it produces sputum and, if so, how much and what kind. Have the patient's cough habits and character of sputum changed recently?

## **PHYSICAL EXAMINATION**

Use inspection skills to check for clues to respiratory disease, beginning with the patient's general appearance. If he or she is frail or cachectic, he or she may have a chronic disease that has impaired his or her appetite. If he or she is diaphoretic, restless, or irritable or protective of a painful body part, he or she may be in acute distress. Also, look for behavior changes that may indicate hypoxemia or hypercapnia. Confusion, lethargy, bizarre behavior, or quiet sleep from which the patient can't be aroused may point to hypercapnia.

Watch for marked cyanosis, indicated by bluish or ashen skin (usually best seen on the lips, tongue, earlobes, and nail beds), which may be due to hypoxemia or poor tissue perfusion.

Assess chest shape and symmetry at rest and during ventilation. Increased anteroposterior diameter (“barrel chest”) characterizes emphysema. Kyphoscoliosis also alters chest configuration, which in turn restricts breathing. Assess respiratory excursion and observe for accessory muscle use during breathing. The use of upper chest and neck muscles is normal only during physical stress.

Observe the rate and pattern of breathing because certain disorders produce characteristic changes in breathing patterns. For example, an acute respiratory disorder can produce tachypnea (rapid, shallow breathing) or hyperpnea (increased rate and depth of breathing); intracranial lesions can produce Cheyne–Stokes and Biot’s respirations; increased intracranial pressure can result in central hyperventilation and apneustic or ataxic breathing; metabolic disorders can cause Kussmaul’s respirations; and airway obstruction can lead to prolonged forceful expiration and pursed-lip breathing.

Also observe posture and carriage. A patient with COPD, for example, usually supports rib cage movement by placing his or her arms on the sides of a chair to increase expansion and lean forward during exhalation to help expel air.

Palpation of the chest wall detects areas of tenderness, masses, changes in fremitus (palpable vocal vibrations), or crepitus (air in subcutaneous tissues). To assess chest excursion and symmetry, place your hands in a horizontal position, bilaterally on the posterior chest, with your thumbs pressed lightly against the spine, creating folds in the skin. As the patient takes a deep breath, your thumbs should move quickly and equally away from the spine. Repeat this with your hands placed anteriorly, at the costal margins (lower lobes) and clavicles (apices). Unequal movement indicates differences in expansion, seen in atelectasis, diaphragm or chest wall muscle disease, or splinting due to pain.

Percussion should detect resonance over lung fields that aren’t covered by bony structures or the heart. A dull sound on percussion may mean consolidation or pleural disease. (See *Characterizing and interpreting percussion sounds*.)

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## Characterizing and Interpreting Percussion Sounds

Percussion may produce several kinds of sounds. Known as flat, dull, resonant, hyperresonant, or tympanic, these sounds indicate the location and density of various structures. During percussion, determining other tonal characteristics, such as pitch, intensity, and quality, also will help identify respiratory structure. Use this chart as a guide to interpreting percussion sounds.

## Characteristic

| <b>Sound</b>   | <b>Pitch</b>         | <b>Intensity</b> | <b>Quality</b>    | <b>Implications</b>  |
|----------------|----------------------|------------------|-------------------|--|
| Flatness       | High                 | Soft             | Extremely dull    | These sounds are normal over the sternum. Over the lung, they may indicate atelectasis or pleural effusion.  |
| Dullness       | Medium               | Medium           | Thudlike          | Normal over the liver, heart, and diaphragm, these sounds over the lung may point to pneumonia, tumor, atelectasis, or pleural effusion.                                 |
| Resonance      | Low                  | Moderate         | Hollow to loud    | When percussed over the lung, these sounds are normal.   |
| Hyperresonance | Lower than resonance | Very loud        | Booming           | These are normal findings with percussion over a child's lung. Over an adult's lung, these findings may indicate emphysema, chronic bronchitis, asthma, or pneumothorax. |
| Tympany        | High                 | Loud             | Musical, drumlike | Over the stomach, these are normal findings; over the lung, they suggest tension pneumothorax.   |

Auscultation normally detects soft, vesicular breath sounds throughout most of the lung fields. Absent or adventitious breath sounds may indicate fluid in small airways or interstitial lung disease (crackles), secretions in moderate and large airways (rhonchi), and airflow obstruction (wheezes).

## SPECIAL RESPIRATORY CARE

The hospitalized patient with respiratory disease may require an artificial upper airway, chest tubes, chest physiotherapy, and supervision of mechanical ventilation. In cardiopulmonary arrest, establishing an airway always takes precedence. In a patient with this condition, airway obstruction usually results when the tongue slides back and blocks the posterior pharynx. The head-tilt method or, in suspected or confirmed cervical fracture or arthritis, the jaw-

thrust maneuver can immediately push the tongue forward, relieving such obstruction. Endotracheal (ET) intubation and, sometimes, a tracheotomy may be necessary.

## CHEST TUBES

An important procedure in patients with respiratory disease is chest tube drainage, which removes air or fluid from the pleural space. This allows the collapsed lung to re-expand to fill the evacuated pleural space. Chest drainage also allows removal of pleural fluid for culture. Chest tubes are commonly used after thoracic surgery, penetrating chest wounds, pleural effusion, and empyema. They're also used for evacuation of pneumothorax, hydrothorax, or hemothorax. Sometimes chest tubes are used to instill sclerosing drugs into the pleural space to prevent recurrent malignant pleural effusions.

Commonly, the chest tube is placed in the sixth or seventh intercostal space, in the axillary region. Occasionally, in pneumothorax, the tube is placed in the second or third intercostal space, in the midclavicular region.

Follow these guidelines when caring for a patient with a chest tube:

- ◆ Monitor changes in suction pressure.
- ◆ Make sure that all connections in the system are tight and secured with tape.
- ◆ Never clamp the chest tube unless checking for air leaks or changing the drainage system.
- ◆ Record the amount, color, and consistency of drainage. Watch for signs of shock, such as tachycardia and hypotension, if drainage is excessive.
- ◆ Encourage the patient to cough and breathe deeply every hour to enhance lung expansion.

Additionally, if a water seal–wet suction system is in place:

- ◆ Check for fluctuation in the water-seal chamber as the patient breathes. Normal fluctuations of 2" to 4" (about 5 to 10 cm) reflect pressure changes in the pleural space during respiration.
- ◆ Watch for intermittent bubbling in the water-seal chamber. This bubbling occurs normally when the system is removing air from the pleural cavity. Absence of bubbling indicates that the pleural space has sealed.
- ◆ Check the water level in the suction-control chamber. If necessary, add sterile water to bring the level to the ordered level.
- ◆ Check for gentle bubbling in the suction-control chamber, which indicates that the proper suction level has been reached.

If a dry-suction system is in place, check that the rotary dry-suction control dial is turned to the ordered suction mark and verify that the appropriate indicator is present, indicating that the desired amount of suction is applied.

## **VENTILATOR METHODS**

Mechanical ventilators are typically used for CNS problems, hypoxemia, or failure of the normal bellows action provided by the diaphragm and rib cage. Positive-pressure ventilators cause inspiration while increasing tidal volume ( $V_T$ ). The inspiratory cycles of these ventilators may vary in volume, pressure, time, or frequency. For example, a volume-cycled ventilator—the type most commonly used—delivers a preset volume of air each time, regardless of the amount of lung resistance. A pressure-cycled ventilator generates flow until the machine reaches a preset pressure regardless of the volume delivered or the time required to achieve the pressure. A time-cycled ventilator generates flow for a preset amount of time. A high-frequency ventilator uses high respiratory rates and low  $V_T$  to maintain alveolar ventilation. Positive end-expiratory pressure (PEEP) is used to retain a certain amount of pressure in the lungs at the end of expiration. By keeping small airways and alveoli open with this method, functional residual capacity is increased and oxygenation is improved.

Implement strategies to prevent ventilator-associated pneumonia (VAP) and plan to remove the patient from ventilator support as soon as the cause of respiratory failure has resolved. (See *Preventing ventilator-associated pneumonia*, page 92.) Several weaning methods are used. The patient may be taken off the ventilator and supplied with a T-piece (ET tube O<sub>2</sub> adapter) that provides O<sub>2</sub> and humidification. The patient then breathes spontaneously without the ventilator for gradually increasing periods.



### **PREVENTION PREVENTING PNEUMONIA**

### **VENTILATOR-ASSOCIATED**

Ventilator-associated pneumonia (VAP) is the leading cause of death among all hospital-acquired infections. VAP also prolongs time spent on the ventilator, length of critical care unit (CCU) stay, and length of hospital stay after discharge from the CCU. Research has shown that the

mortality rate due to VAP can be reduced by early recognition of pneumonia and consistent application of evidence-based practices. The Ventilator Bundle is a group of interventions related to ventilator care that, when implemented together, achieve significantly better outcomes than when implemented individually. The key components of the Ventilator Bundle include:

- ◆ elevating the head of the bed 30 to 45 degrees
- ◆ interrupting sedation daily and assessing the readiness to extubate
- ◆ instituting peptic ulcer disease prophylaxis
- ◆ instituting deep vein thrombosis prophylaxis
- ◆ providing daily oral care with chlorhexidine

Various other best practices can be combined with the bundle to prevent VAP. They include:

- ◆ adhering to Centers for Disease Control and Prevention or World Health Organization hand hygiene guidelines to prevent the spread of infection
- ◆ using noninvasive ventilatory support, such as bilevel positive-airway ventilation instead of endotracheal (ET) intubation and mechanical ventilation, to eliminate the risk of VAP
- ◆ using the oral route instead of the nasal route for ET intubation to prevent sinusitis
- ◆ maintaining ET tube cuff pressure at 20 cm or more to prevent aspiration
- ◆ using a cuffed ET tube with in-line and subglottic suctioning to prevent secretion aspiration
- ◆ avoiding gastric distention to reduce the risk for aspiration
- ◆ avoiding unexplained extubation and reintubation to prevent secretion aspiration
- ◆ minimizing equipment contamination (by removing condensate from ventilator circuits, keeping the circuit closed during removal, changing the ventilator circuit only when visibly soiled or malfunctioning, and disinfecting and storing respiratory equipment properly) to prevent airway contamination
- ◆ teaching the patient and family about measures to prevent VAP and involving them in monitoring

With intermittent mandatory ventilation, the ventilator provides a specific number of breaths, and the patient is able to breathe spontaneously between ventilator breaths. The frequency of ventilator breaths is gradually decreased until the patient can breathe on his or her own. Pressure support ventilation, in which the patient receives a preset pressure boost with each spontaneous breath, has proved effective. Vital signs, ABG levels, physical findings, and subjective symptoms should be monitored periodically during weaning to assess respiratory status.

## Chest Physiotherapy

In respiratory conditions marked by excessive accumulation of secretions in the lungs, chest physiotherapy may enhance removal of secretions. Chest physiotherapy includes chest assessment, effective breathing and coughing exercises, postural drainage, percussion, vibration, and evaluation of the therapy's effectiveness. Before initiating treatment, review X-rays and physical assessment findings to locate areas of secretions.

- ◆ Deep breathing maintains diaphragm use, increases negative intrathoracic pressure, and promotes venous return; it's especially important when pain or dressings restrict chest movement. An incentive spirometer can provide positive visual reinforcement to promote deep breathing.
- ◆ Pursed-lip breathing is used primarily in obstructive disease to slow expiration and prevent small airway collapse. Such breathing slows air through smaller bronchi, maintaining positive pressure and preventing collapse of small airways and resultant air trapping.
- ◆ Segmental breathing or lateral costal breathing is used after lung resection and for localized disorders. Place your hand over the lung area on the affected side. Instruct the patient to try to push that portion of the chest against your hand on deep inspiration. You should be able to feel this with your hand.
- ◆ Coughing that's controlled and staged gradually increases intrathoracic pressure, reducing pain and bronchospasm of explosive coughing. When wound pain prevents effective coughing, splint the wound with a pillow, towel, or your hand during coughing exercises.
- ◆ Postural drainage uses gravity to drain secretions into larger airways, where they can be expectorated. This technique is used in the patient with copious or tenacious secretions. Before performing postural drainage, auscultate the patient's chest and review chest X-rays to determine the best

position for maximum drainage. To prevent vomiting, schedule postural drainage at least 1 hour after meals.

- ◆ Percussion moves air against the chest wall, enhancing the effectiveness of postural drainage by loosening lung secretions. Percussion is contraindicated in severe pain, extreme obesity, cancer that has metastasized to the ribs, crushing chest injuries, bleeding disorders, spontaneous pneumothorax, spinal compression fractures, and in patients with temporary pacemakers.
- ◆ Vibration can be used with percussion or alone when percussion is contraindicated.
- ◆ PEEP therapy maintains positive pressure in airways, preventing small airway collapse.

Before and after chest physiotherapy, auscultate the patient's lung fields and assess for sputum production to evaluate the effectiveness of therapy.

## Congenital and Pediatric Disorders

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### RESPIRATORY DISTRESS SYNDROME

Respiratory distress syndrome (RDS), also called *hyaline membrane disease*, is the most common cause of neonatal mortality. In the United States alone, it kills 40,000 neonates every year. RDS occurs in premature neonates and, if untreated, is fatal within 72 hours of birth in up to 14% of neonates weighing less than  $5\frac{1}{2}$  lb (2.5 kg). Aggressive management using mechanical ventilation can improve the prognosis, but some surviving neonates may develop some degree of bronchopulmonary dysplasia (BPD).

#### Causes and Incidence

Although airways and alveoli of a neonate's respiratory system are present by 27 weeks' gestation, the intercostal muscles are weak, and the alveolar capillary system is immature. The preterm neonate with RDS develops widespread alveolar collapse because of a lack of surfactant, a lipoprotein present in alveoli and respiratory bronchioles. The surfactant lowers surface tension and helps prevent alveolar collapse. This surfactant deficiency results in widespread atelectasis, which leads to inadequate alveolar ventilation with shunting of blood through collapsed areas of lung, causing hypoxemia and acidosis.

RDS occurs almost exclusively in neonates born before 37 weeks' gestation (in 60% of those born before the 28th week). The incidence is greatest in those with birth weights of 1,000 to 1,500 g. Infants of diabetic mothers, those born by cesarean delivery, second-born twins, infants with perinatal asphyxia, and those delivered suddenly after antepartum hemorrhage are more commonly affected.

## Pathophysiology

The lack of surfactant coating the alveoli reduces the available pulmonary surface and decreases the area for gas exchange. Worsening hypercapnia and hypoxia cause metabolic and respiratory acidosis leading to pulmonary vasoconstriction and peripheral vasodilation. Damage to the endothelial and alveolar cells results from the ongoing hypoxia. The subsequent vascular disruption leads to plasma leakage into the alveolar spaces, layering of fibrin and necrotic cells creating hyaline membranes. These membranes impede the exchange of gases across the alveolar surface.

## Complications

- ◆ Pneumothorax
- ◆ Pneumomediastinum
- ◆ Pneumopericardium
- ◆ BPD
- ◆ Intraventricular bleed
- ◆ Hemorrhage into lungs after surfactant use
- ◆ Retinopathy of prematurity (or ROP)
- ◆ Delayed mental development or mental retardation

## Signs and Symptoms

Although a neonate with RDS may breathe normally at first, they usually develop rapid, shallow respirations within minutes or hours of birth, with intercostal, subcostal, or sternal retractions; nasal flaring; and audible expiratory grunting. This grunting is a natural compensatory mechanism designed to produce PEEP and prevent further alveolar collapse.

Severe disease is marked by apnea, bradycardia, and cyanosis (from hypoxemia, left-to-right shunting through the foramen ovale, or right-to-left intrapulmonary shunting through atelectatic regions of the lung). Other

clinical features include pallor, frothy sputum, and low body temperature as a result of an immature nervous system and the absence of subcutaneous fat.

## Diagnosis

**Dx CONFIRMING DIAGNOSIS** *Signs of respiratory distress in a premature neonate during the first few hours of life strongly suggest RDS, but a chest X-ray and ABG analysis are needed to confirm the diagnosis.*

- ◆ Chest X-ray may be normal for the first 6 to 12 hours (in 50% of neonates with RDS), but 24 hours after birth it will show the characteristic ground-glass appearance and air bronchograms.
- ◆ ABG analysis shows decreased  $\text{Pao}_2$ ; normal, decreased, or increased  $\text{Paco}_2$ ; and decreased pH (from respiratory or metabolic acidosis or both).
- ◆ Chest auscultation reveals normal or diminished air entry and crackles (rare in early stages).

When a cesarean birth is necessary before 36 weeks' gestation, amniocentesis enables the determination of the lecithin/sphingomyelin (L/S) ratio and the presence of phosphatidylglycerol. An L/S ratio of more than 2:1 and the presence of phosphatidylglycerol decrease the likelihood of RDS.

## Treatment

Treatment of an infant with RDS requires vigorous respiratory support. Warm, humidified, oxygen-enriched gases are administered by oxygen hood or, if such treatment fails, by mechanical ventilation. Severe cases may require mechanical ventilation with PEEP or continuous positive-airway pressure (CPAP), administered by nasal prongs or, when necessary, ET intubation. Special ventilation techniques are now used on the patient's refractory to conventional mechanical ventilation. These include high-frequency jet ventilation and high-frequency oscillatory ventilation. Extracorporeal membrane oxygenation is the last choice for ventilation and is only available in certain specialized facilities. Treatment of RDS also includes:

- ◆ a radiant warmer or isolette for thermoregulation
- ◆ I.V. fluids and sodium bicarbonate to control acidosis and maintain fluid and electrolyte balance
- ◆ tube feedings or total parenteral nutrition if the neonate is too weak to eat

- ◆ administration of surfactant by an ET tube (Studies show that this treatment can prevent or improve the course of RDS as well as reduce mortality.)

## Special Considerations

- ◆ Neonates with RDS require continual assessment and monitoring in an intensive care nursery.
- ◆ Closely monitor ABGs as well as fluid intake and output. If the neonate has an umbilical catheter (arterial or venous), check for arterial hypotension or abnormal central venous pressure. Watch for complications, such as infection, thrombosis, or decreased circulation to the legs. If the neonate has a transcutaneous oxygen monitor, change the site of the lead placement every 2 to 4 hours.
- ◆ To evaluate progress, assess skin color, rate and depth of respirations, severity of retractions, nostril flaring, frequency of expiratory grunting, frothing at the lips, and restlessness.
- ◆ Regularly assess the effectiveness of oxygen or ventilator therapy. Evaluate every change in fraction of inspired oxygen and PEEP or CPAP by monitoring arterial oxygen saturation or ABG levels. Adjust the PEEP or CPAP as indicated, on the basis of findings.
- ◆ Mechanical ventilation in neonates is usually done in a pressure-limited mode rather than in the volume-limited mode used in adults.
- ◆ When the neonate is on mechanical ventilation, watch carefully for signs of barotrauma (an increase in respiratory distress and subcutaneous emphysema) and accidental disconnection from the ventilator. Check ventilator settings frequently. Be alert for signs of complications of PEEP or CPAP therapy, such as decreased cardiac output, pneumothorax, and pneumomediastinum. Mechanical ventilation increases the risk of infection in the preterm neonate, so preventive measures are essential.
- ◆ As needed, arrange for follow-up care with a neonatal ophthalmologist to check for retinal damage. Preterm neonates in an oxygen-rich environment are at increased risk for developing ROP.
- ◆ Teach the parents about their neonate's condition and, if possible, let them participate in their care (using sterile technique), to encourage normal parent–infant bonding. Advise parents that full recovery may take up to 12 months. When the prognosis is poor, prepare the parents for the neonate's impending death and offer emotional support.

- ◆ Help reduce mortality in the neonate with RDS by detecting respiratory distress early. Recognize intercostal retractions and grunting, especially in a premature neonate, as signs of RDS; make sure the neonate receives immediate treatment.



## PREVENTION

- ◆ *Prenatal care can help prevent prematurity.*
- ◆ *Give corticosteroids to the mother 2 to 3 days before delivery to help the infant's lungs mature in preterm deliveries.*

## SUDDEN INFANT DEATH SYNDROME

A medical mystery of early infancy, sudden infant death syndrome (SIDS), also called *crib death*, is the unexpected, sudden death of an infant or child younger than age 1 year. Reasons for the death remain unexplained even after an autopsy. Typically, parents put the infant to bed and later find him or her dead, commonly with no indications of a struggle or distress of any kind. Incidence has decreased with the practice of teaching parents to place an infant on their back to sleep.

### Causes and Incidence

SIDS is the third leading cause of death in infants between 1 month and 1 year old. It occurs more commonly in winter months. The incidence is higher in males, preterm neonates, and those who sleep on their stomachs or in cribs with soft bedding. Incidence is also higher among neonates born in conditions of poverty and to those who were one of a single multiple birth, such as twins and triplets, and to mothers who smoke, take drugs, or failed to seek prenatal care until late in the pregnancy. SIDS may also result from an abnormality in the control of ventilation that allows CO<sub>2</sub> to build up in the blood, thereby causing prolonged apneic periods with profound hypoxemia and serious cardiac arrhythmias. It's also thought to be associated with problems in sleep arousal.

### Pathophysiology

Although the exact pathophysiology of SIDS is not known, there is a common theory. Abnormalities of the autoimmune nervous system and brainstem cause dysfunctions of breathing. Episodes of hypoxia contribute to delaying the

arousal response when oxygen availability is decreased and potentially leading to death.

## **Signs and Symptoms**

Although parents find some victims wedged in crib corners or with blankets wrapped around their heads, autopsies rule out suffocation as the cause of death. Autopsy shows a patent airway, so aspiration of vomitus isn't the cause of death. Typically, SIDS babies don't cry out and show no signs of having been disturbed in their sleep. However, their positions or tangled blankets may suggest movement just before death, perhaps due to terminal spasm.

Depending on how long the infant has been dead, a SIDS baby may have a mottled complexion with extreme cyanosis of the lips and fingertips or pooling of blood in the legs and feet that may be mistaken for bruises. Pulse and respirations are absent, and the diaper is wet and full of stool.

## **Diagnosis**

Diagnosis of SIDS requires an autopsy to rule out other causes of death. Characteristic histologic findings on autopsy include small or normal adrenal glands and petechiae over the visceral surfaces of the pleura, within the thymus, and in the epicardium. Autopsy also reveals extremely well-preserved lymphoid structures and certain pathologic characteristics that suggest chronic hypoxemia such as increased pulmonary artery smooth muscle. Examination also shows edematous, congestive lungs fully expanded in the pleural cavities, liquid (not clotted) blood in the heart, and curd from the stomach inside the trachea.

## **Treatment**

If the parents bring the infant to the emergency department (ED), the physician will decide whether to try to resuscitate him. An "aborted SIDS" infant is one who's found apneic and is successfully resuscitated. Such an infant, or any infant who had a sibling stricken by SIDS, should be tested for infantile apnea. If tests are positive, a home apnea monitor may be recommended. Because the infant usually can't be resuscitated, however, treatment focuses on providing emotional support for the family.

## **Special Considerations**

- ◆ Make sure that parents are present when the child's death is announced. They may lash out at ED personnel, the babysitter, or anyone else involved in the child's care—even each other. Stay calm and let them express their feelings. Reassure them that they weren't to blame.
- ◆ Let the parents see the baby in a private room. Allow them to express their grief in their own way. Stay in the room with them if appropriate. Offer to call clergy, friends, or relatives.
- ◆ After the parents and family have recovered from their initial shock, explain the necessity for an autopsy to confirm the diagnosis of SIDS (in some states, this is mandatory). At this time, provide the family with some basic facts about SIDS and encourage them to give their consent for the autopsy. Make sure that they receive the autopsy report promptly.
- ◆ Find out whether your community has a local counseling and information program for SIDS parents. Participants in such a program will contact the parents, ensure that they receive the autopsy report promptly, put them in touch with a professional counselor, and maintain supportive telephone contact. Also, find out whether there's a local SIDS parent group; such a group can provide significant emotional support. Contact the National Sudden Infant Death Foundation for information about such local groups.
- ◆ If your facility's policy is to assign a public health nurse to the family, they will provide the continuing reassurance and assistance the parents will need.
- ◆ If the parents decide to have another child, they'll need information and counseling to help them through the pregnancy and the first year of the new infant's life.
- ◆ Infants at high risk for SIDS may be placed on apnea monitoring at home.
- ◆ All new parents should be informed of the American Academy of Pediatrics' recommendation that infants be positioned on their back, not on their stomach or side, for sleeping.



## **PREVENTION**

- ◆ *Tell parents to place infants on their backs to sleep.*
- ◆ *Tell parents infants should sleep on a firm mattress and shouldn't have soft objects in the crib; like stuffed toys and blankets.*
- ◆ *Tell parents infants shouldn't sleep in the same bed as their parents.*
- ◆ *Tell parents to give infants pacifiers at bedtime.*
- ◆ *Tell parents infants shouldn't be exposed to secondhand smoke.*

## CROUP

Croup is a severe inflammation and obstruction of the upper airway, occurring as acute laryngotracheobronchitis (most common), laryngitis, and acute spasmodic laryngitis; it must always be distinguished from epiglottitis. It's derived from an old German word for "voice box" and refers to swelling around the larynx or vocal cords. Recovery is usually complete.

### Causes and Incidence

Croup usually results from a viral infection but can also be caused by bacteria, allergens, and inhaled irritants. Parainfluenza viruses cause 75% of such infections; adenoviruses, respiratory syncytial virus (RSV), influenza, and measles viruses account for the rest.

Croup is a childhood disease affecting more boys than girls (typically between 3 months and 5 years old) that usually occurs during the winter. Up to 15% of patients have a strong family history of croup.

### Pathophysiology

Infection of the laryngeal mucosa leads to edema and inflammation of the epiglottal area. This swelling leads to a narrowing of the airway and increasingly deep respirations. The ongoing effort to breath as the narrowing progresses becomes more difficult and the air flowing through the upper airway becomes turbulent. During inspiration, the flexible chest wall caves in slightly and causing paradoxical breathing.

### Complications

- ◆ Respiratory distress
- ◆ Respiratory arrest
- ◆ Epiglottitis
- ◆ Bacterial tracheitis
- ◆ Atelectasis
- ◆ Dehydration

### Signs and Symptoms

The onset of croup usually follows an upper respiratory tract infection. Clinical features include inspiratory stridor, hoarse or muffled vocal sounds, varying degrees of laryngeal obstruction and respiratory distress, and a characteristic sharp, barking, seal-like cough. These symptoms may last only

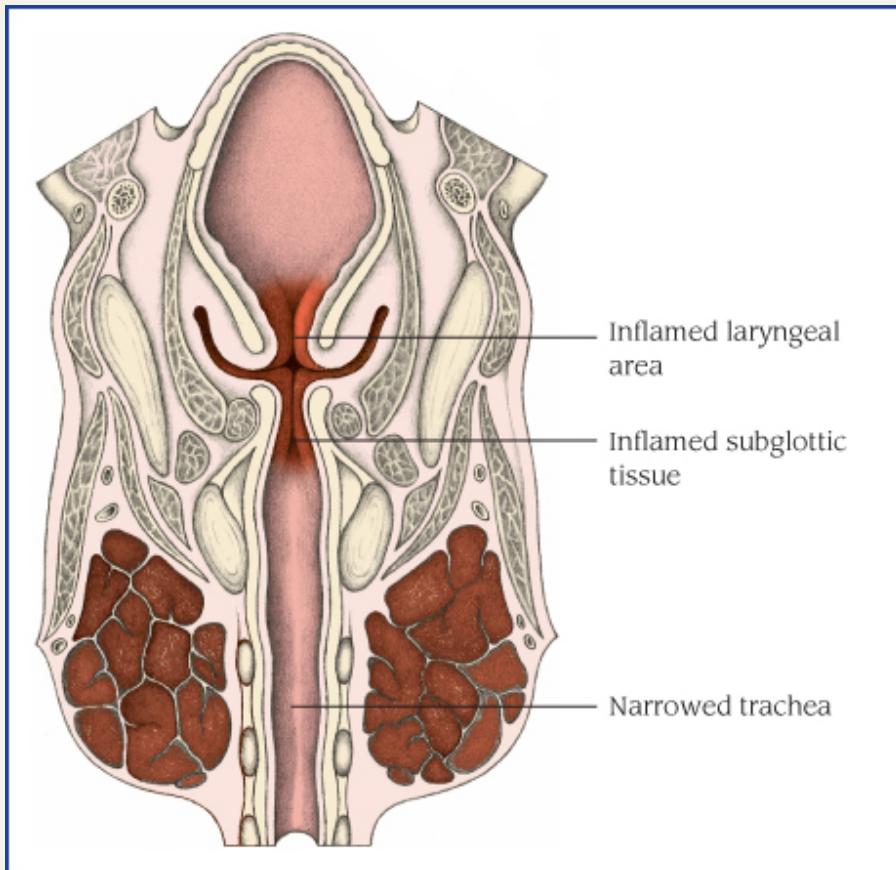
a few hours or persist for a day or two. As it progresses, croup causes inflammatory edema and, possibly, spasm, which can obstruct the upper airway and severely compromise ventilation. (See *How croup affects the upper airway*.)



## PATHOPHYSIOLOGY

### HOW CROUP AFFECTS THE UPPER AIRWAY

In croup, inflammatory swelling and spasms constrict the larynx, thereby reducing airflow. This cross-sectional drawing (from chin to chest) shows the upper airway changes caused by croup. Inflammatory changes almost completely obstruct the larynx (which includes the epiglottis) and significantly narrow the trachea.



Each form of croup has additional characteristics:

In *laryngotracheobronchitis*, the symptoms seem to worsen at night. Inflammation causes edema of the bronchi and bronchioles as well as increasingly difficult expiration that frightens the child. Other characteristic features include fever, diffusely decreased breath sounds, expiratory rhonchi, and scattered crackles.

*Laryngitis*, which results from vocal cord edema, is usually mild and produces no respiratory distress except in infants. Early signs include a sore throat and cough, which, rarely, may progress to marked hoarseness, suprasternal and intercostal retractions, inspiratory stridor, dyspnea, diminished breath sounds, restlessness and, in later stages, severe dyspnea and exhaustion.

*Acute spasmodic laryngitis* affects a child between 1 and 3 years old, particularly one with allergies and a family history of croup. It typically begins with mild to moderate hoarseness and nasal discharge, followed by the characteristic cough and noisy inspiration (that usually awaken the child at night), labored breathing with retractions, rapid pulse, and clammy skin. The child understandably becomes anxious, which may lead to increasing dyspnea and transient cyanosis. These severe symptoms diminish after several hours but reappear in a milder form on the next one or two nights.

## Diagnosis

The clinical picture is very characteristic, so the diagnosis should be suspected immediately. When bacterial infection is the cause, throat cultures may identify the organisms and their sensitivity to antibiotics and rule out diphtheria. On a posterior–anterior X-ray of the chest, narrowing of the upper airway (“steeple sign”) may be apparent. Laryngoscopy may reveal inflammation and obstruction in epiglottal and laryngeal areas. In evaluating the patient, assess for foreign body obstruction (a common cause of crouplike cough in a young child) as well as masses and cysts.

## Treatment

For most children with croup, home care with rest, cool mist humidification during sleep, and antipyretics, such as acetaminophen, relieve symptoms. However, respiratory distress that’s severe or interferes with oral hydration requires hospitalization and parenteral fluid replacement to prevent dehydration. If bacterial infection is the cause, antibiotic therapy is necessary. Oxygen therapy may also be required. Increasing obstruction of the airway requires intubation and mechanical ventilation.

Inhaled racemic epinephrine and corticosteroids may be used to alleviate respiratory distress.

## Special Considerations

Monitor and support respiration, and control fever. Because croup is so frightening to the child and family, you must also provide support and reassurance.

- ◆ Carefully monitor cough and breath sounds, hoarseness, severity of retractions, inspiratory stridor, cyanosis, respiratory rate and character (especially prolonged and labored respirations), restlessness, fever, and cardiac rate.
- ◆ Keep the child as quiet as possible. However, avoid sedation because it may depress respiration. If the patient is an infant, position them in an infant seat or propped up with a pillow; place an older child in Fowler's position. If an older child requires a cool mist tent to help them breathe, explain why it's needed.
- ◆ Isolate patients suspected of having RSV and parainfluenza infections if possible. Wash your hands carefully before leaving the room, to avoid transmission to other children, particularly infants. Instruct parents and others involved in the care of these children to take similar precautions.
- ◆ Control fever with sponge baths and antipyretics. Keep a hypothermia blanket on hand for temperatures above 102° F (38.9° C). Watch for seizures in infants and young children with high fevers. Give I.V. antibiotics as ordered.
- ◆ Relieve sore throat with soothing, water-based ices, such as fruit sherbet and ice pops. Avoid thicker, milk-based fluids if the child is producing heavy mucus or has great difficulty in swallowing. Apply petroleum jelly or another ointment around the nose and lips to soothe irritation from nasal discharge and mouth breathing.
- ◆ Maintain a calm, quiet environment and offer reassurance. Explain all procedures and answer any questions.

When croup doesn't require hospitalization:

- ◆ Teach the parents effective home care. Suggest the use of a cool mist humidifier (vaporizer). To relieve croupy spells, tell parents to carry the child into the bathroom, shut the door, and turn on the hot water. Breathing in warm, moist air quickly eases an acute spell of croup.

- ◆ Warn parents that ear infections and pneumonia are complications of croup, which may appear about 5 days after recovery. Stress the importance of immediately reporting earache, productive cough, high fever, or increased shortness of breath.



## PREVENTION

- ◆ *Perform hand hygiene frequently to prevent a respiratory infection.*
- ◆ *Give diphtheria, tetanus, and pertussis (DpT); Haemophilus influenzae B (Hib); and measles, mumps, and rubella (MMR) vaccines to children.*

## EPIGLOTTITIS

Acute epiglottitis is an acute inflammation of the epiglottis that tends to cause airway obstruction. A critical emergency, epiglottitis can prove fatal unless it's recognized and treated promptly.

### Causes and Incidence

Epiglottitis usually results from infection with Hib and, occasionally, pneumococci and group A streptococci. It typically strikes children between 2 and 6 years old. (However, immunosuppression can predispose adults to epiglottitis.) Since the advent of the Hib vaccine, epiglottitis is becoming more rare.

### Pathophysiology

The causative bacteria invade the mucosa and into the bloodstream causing bacteremia and infection of the epiglottis as well as surrounding tissues. Acute inflammation and edema begin in the epiglottic area and progressing to the epiglottic folds, arytenoids, and entire supraglottic larynx. The aggressive swelling and edema greatly reduces the available airway and quickly increasing the risk for a respiratory crisis.

### Complications

- ◆ Respiratory failure
- ◆ Pneumonia
- ◆ Meningitis
- ◆ Death
- ◆ Pericarditis

## **Signs And Symptoms**

Sometimes preceded by an upper respiratory infection, epiglottitis may rapidly progress to complete upper airway obstruction within 2 to 5 hours. Laryngeal obstruction results from inflammation and edema of the epiglottis. Accompanying symptoms include high fever, stridor, sore throat, dysphagia, irritability, restlessness, and drooling. To relieve severe respiratory distress, the child with epiglottitis may hyperextend his or her neck, sit up, and lean forward with his or her mouth open, tongue protruding, and nostrils flaring as he or she tries to breathe. The child may develop inspiratory retractions and rhonchi.

## **Diagnosis**

In acute epiglottitis, throat examination reveals a large, edematous, bright red epiglottis. Such examination should follow lateral neck X-rays and, generally, *shouldn't* be performed if the suspected obstruction is great. Special equipment (laryngoscope and ET tubes) should be available because a tongue blade can cause sudden complete airway obstruction. Trained personnel (such as an anesthesiologist) should be on hand during the throat examination to secure an emergency airway. On the lateral soft-tissue X-ray of the neck, a large, thick but indistinct ("thumbprint") epiglottis will be seen. Blood or throat culture may show *H. influenzae* or other bacteria.

## **Treatment**

A child with acute epiglottitis and airway obstruction requires emergency hospitalization; the child may need emergency ET intubation or a tracheotomy with subsequent monitoring in an intensive care unit. Respiratory distress that interferes with swallowing necessitates parenteral fluid administration to prevent dehydration. A patient with acute epiglottitis should always receive a complete course of parenteral antibiotics—usually a second- or third-generation cephalosporin. (If the child is allergic to penicillin, a quinolone or sulfa drug may be substituted.) Corticosteroids should be used to decrease swelling of the throat.

## **Special Considerations**

- ◆ Keep equipment available in case of sudden complete airway obstruction to secure an airway. Be prepared to assist with intubation or tracheotomy, as necessary.



**ALERT** *Watch for increasing restlessness, rising cardiac rate, fever, dyspnea, and retractions, which may indicate the need for an emergency tracheotomy. Monitor blood gases for hypoxemia and hypercapnia.*

- ◆ After a tracheotomy, anticipate the patient's needs because they won't be able to cry or call out; provide emotional support. Reassure the patient and their family that the tracheotomy is a short-term intervention (usually from 4 to 7 days). Monitor the patient for rising temperature and pulse rate and hypotension—signs of secondary infection.
- ◆ The bacterial infection causing epiglottitis is contagious, and airborne or droplet precautions should be followed. Family members should be screened.



### PREVENTION

- ◆ *Perform hand hygiene frequently to prevent infections.*
- ◆ *Administer the Hib vaccine to children.*

## Acute Disorders

### ACUTE RESPIRATORY DISTRESS SYNDROME

A form of noncardiogenic pulmonary edema that causes acute respiratory failure (ARF), acute respiratory distress syndrome (ARDS), also called *shock lung* or *adult respiratory distress syndrome*, results from increased permeability of the alveolocapillary membrane. Fluid accumulates in the lung interstitium, alveolar spaces, and small airways, causing the lung to stiffen. Effective ventilation is thus impaired, prohibiting adequate oxygenation of pulmonary capillary blood. Severe ARDS can cause intractable and fatal hypoxemia. However, patients who recover may have little or no permanent lung damage. (See *Alveolar changes in ARDS*.)



### PATHOPHYSIOLOGY ALVEOLAR CHANGES IN ARDS

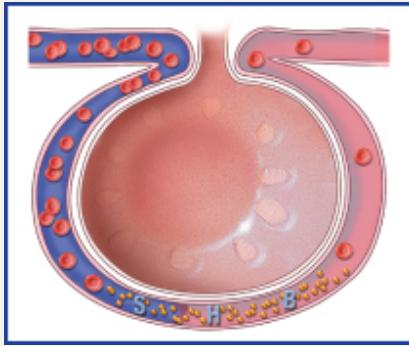
The alveoli undergo major changes in each phase of ARDS.

Phase 1

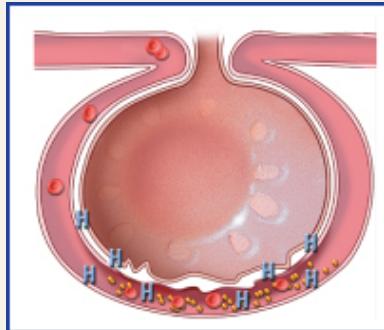
Phase 2

Phase 3

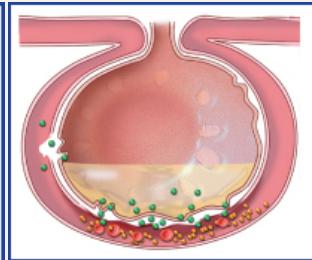
In phase 1, injury reduces normal blood flow to the lungs. Platelets aggregate and release histamine (H), serotonin (S), and bradykinin (B).



In phase 2, those substances—especially histamine—inflame and damage the alveolocapillary membrane, increasing capillary permeability. Fluids then shift into the interstitial

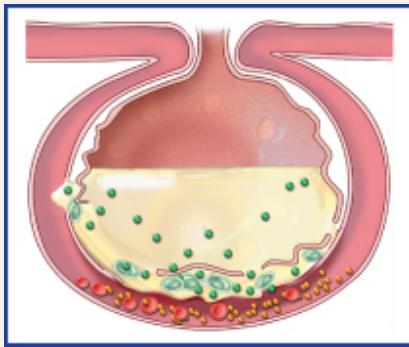


In phase 3, as capillary permeability increases, proteins and fluids leak out, increasing interstitial osmotic pressure and causing pulmonary edema.



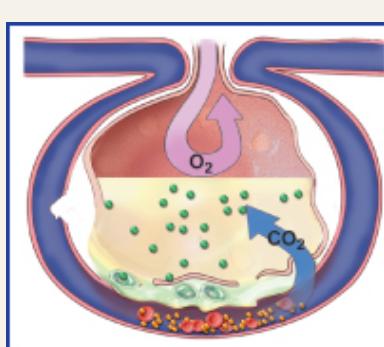
#### Phase 4

In phase 4, decreased blood flow and fluids in the alveoli damage surfactant and impair the cell's ability to produce more. As a result, alveoli collapse, impeding gas exchange and decreasing lung compliance.



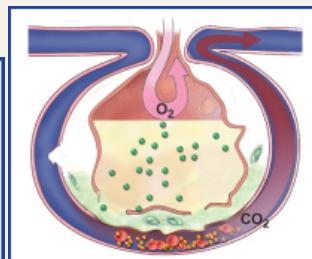
#### Phase 5

In phase 5, sufficient oxygen can't cross the alveolocapillary membrane, but carbon dioxide inflammation leads to ( $\text{CO}_2$ ) can and is lost with every exhalation. Oxygen ( $\text{O}_2$ ) levels decrease in the blood.



#### Phase 6

In phase 6, pulmonary fibrosis, and gas exchange is further impeded.



## Causes and Incidence

ARDS results from many respiratory and nonrespiratory insults, such as:

- ◆ aspiration of gastric contents
- ◆ sepsis (primarily gram-negative), trauma, or oxygen toxicity
- ◆ shock

- ◆ viral, bacterial, or fungal pneumonia or microemboli (fat or air emboli or disseminated intravascular coagulation)
- ◆ drug overdose (barbiturates, glutethimide, or opioids)
- ◆ blood transfusion
- ◆ smoke or chemical inhalation (nitrous oxide, chlorine, or ammonia)
- ◆ hydrocarbon and paraquat ingestion
- ◆ pancreatitis, uremia, or miliary tuberculosis (TB; rare)
- ◆ near drowning
- ◆ severe traumatic injuries, such as head injury or pulmonary contusions

Altered permeability of the alveolocapillary membrane causes fluid to accumulate in the interstitial space. If the pulmonary lymphatic glands can't remove this fluid, interstitial edema develops. The fluid collects in the peribronchial and peribronchiolar spaces, producing bronchiolar narrowing. Hypoxemia occurs as a result of fluid accumulation in alveoli and subsequent alveolar collapse, causing the shunting of blood through nonventilated lung regions. In addition, alveolar collapse causes a dramatic increase in lung compliance, which makes it more difficult to achieve adequate ventilation.

ARDS affects 10 to 14 people per 100,000, with a mortality rate of 36% to 52%.

## **Pathophysiology**

An acute lung injury can begin the cascade of alveolar damage resulting in an altered permeability of the epithelial barrier and subsequent pulmonary edema. Diffuse alveolar damage progresses and granulation tissue forms in the alveolar spaces creating fibrosis. This fibrotic phase inhibits lung compliance and effective respiration.

## **Complications**

- ◆ Multisystem failure
- ◆ Pulmonary fibrosis
- ◆ Pneumothorax

## **Signs and Symptoms**

ARDS initially produces rapid, shallow breathing and dyspnea within hours to days of the initial injury (sometimes after the patient's condition appears to have stabilized). Hypoxemia develops, causing an increased drive for ventilation. Because of the effort required to expand the stiff lung, intercostal

and suprasternal retractions result. Fluid accumulation produces crackles and rhonchi; worsening hypoxemia causes restlessness, apprehension, mental sluggishness, motor dysfunction, and tachycardia (possibly with transient increased arterial blood pressure).



**ELDER TIP** *The older patient may appear to do well following an initial episode of ARDS. Symptoms commonly appear 2 to 3 days later.*

Severe ARDS causes overwhelming hypoxemia. If uncorrected, this results in hypotension, decreasing urine output, respiratory and metabolic acidosis, and eventually ventricular fibrillation or standstill.

## Diagnosis

On room air, ABG analysis initially shows decreased Pao<sub>2</sub> (less than 60 mm Hg) and Paco<sub>2</sub> (less than 35 mm Hg). The resulting pH usually reflects respiratory alkalosis. As ARDS becomes more severe, ABG analysis shows respiratory acidosis (increasing Paco<sub>2</sub> [more than 45 mm Hg]), metabolic acidosis (decreasing bicarbonate [less than 22 mEq/L]), and a decreasing Pao<sub>2</sub>, despite oxygen therapy.

Other diagnostic tests include the following:

- ◆ Pulmonary artery catheterization helps identify the cause of pulmonary edema (cardiac versus noncardiac) by evaluating pulmonary artery wedge pressure (PAWP); allows collection of pulmonary artery blood, which shows decreased oxygen saturation, reflecting tissue hypoxia; measures pulmonary artery pressure (PAP); measures cardiac output by thermodilution techniques; and provides information to allow calculation of the percentage of blood shunted through the lungs.
- ◆ Serial chest X-rays initially show bilateral infiltrates. In later stages, a ground-glass appearance and eventually (as hypoxemia becomes irreversible), “whiteouts” of both lung fields are apparent. Medical personnel can differentiate ARDS from heart failure by noting the following on serial chest X-rays:
  - ◆ normal cardiac silhouette
  - ◆ diffuse bilateral infiltrates that tend to be more peripheral and patchy, as opposed to the usual perihilar “bat wing” appearance of cardiogenic pulmonary edema
  - ◆ fewer pleural effusions

Differential diagnosis must rule out cardiogenic pulmonary edema, pulmonary vasculitis, and diffuse pulmonary hemorrhage. To establish the etiology, laboratory work should include sputum Gram stain, culture and sensitivity tests, and blood cultures to detect infections; a toxicology screen for drug ingestion; and, when pancreatitis is a consideration, a serum amylase determination.

## Treatment

When possible, treatment is designed to correct the underlying cause of ARDS as well as to prevent progression and the potentially fatal complications of hypoxemia and respiratory acidosis. Supportive medical care consists of administering humidified oxygen with CPAP. Hypoxemia that doesn't respond adequately to these measures requires ventilatory support with intubation, volume ventilation, and PEEP. Other supportive measures include fluid restriction, diuretics, and correction of electrolyte and acid–base abnormalities.

When ARDS requires mechanical ventilation, sedatives, opioids, or neuromuscular blocking agents may be ordered to optimize ventilation. Treatment to reverse severe metabolic acidosis with sodium bicarbonate may be necessary, although in severe cases this may worsen the acidosis if CO<sub>2</sub> can't be cleared adequately. Use of fluids and vasopressors may be required to maintain blood pressure. Infections require appropriate anti-infective therapy.

## Special Considerations

ARDS requires careful monitoring and supportive care.

- ◆ Frequently assess the patient's respiratory status. Be alert for retractions on inspiration. Note the rate, rhythm, and depth of respirations; watch for dyspnea and the use of accessory muscles of respiration. On auscultation, listen for adventitious or diminished breath sounds. Check for clear, frothy sputum, which may indicate pulmonary edema.
- ◆ Observe and document the hypoxic patient's neurologic status (level of consciousness and mental status).
- ◆ Maintain a patent airway by suctioning, using sterile, nontraumatic technique. Ensure adequate humidification to help liquefy tenacious secretions.
- ◆ Closely monitor heart rate and blood pressure. Watch for arrhythmias that may result from hypoxemia, acid–base disturbances, or electrolyte

imbalance. With pulmonary artery catheterization, know the desired pressure levels. Check readings often and watch for decreasing mixed venous oxygen saturation.

- ◆ Monitor serum electrolytes and correct imbalances. Measure intake and output; weigh the patient daily.
- ◆ Check ventilator settings frequently, and empty condensate from tubing promptly to ensure maximum oxygen delivery. Monitor ABG studies and pulse oximetry. The patient with severe hypoxemia may need controlled mechanical ventilation with positive pressure. Give sedatives, as needed, to reduce restlessness.
- ◆ Because PEEP may decrease cardiac output, check for hypotension, tachycardia, and decreased urine output. Suction only as needed to maintain PEEP or use an in-line suctioning apparatus. Reposition the patient often and record an increase in secretions, temperature, or hypotension that may indicate a deteriorating condition. Monitor peak pressures during ventilation. Because of stiff, noncompliant lungs, the patient is at high risk for barotrauma (pneumothorax), evidenced by increased peak pressures, decreased breath sounds on one side, and restlessness.
- ◆ Monitor nutrition, maintain joint mobility, and prevent skin breakdown. Accurately record calorie intake. Give tube feedings and parenteral nutrition, as ordered. Perform passive range-of-motion exercises or help the patient perform active exercises, if possible. Provide meticulous skin care. Plan patient care to allow periods of uninterrupted sleep.
- ◆ Provide emotional support. Warn the family and the patient who's recovering from ARDS that recovery will take some time and that they will feel weak for a while.
- ◆ Watch for and immediately report all respiratory changes in the patient with injuries that may adversely affect the lungs (especially during the 2- to 3-day period after the injury, when the patient may appear to be improving).



**PREVENTION** Prevent VAP through use of the Ventilator Bundle and other best practices, such as continuous removal of subglottic secretions, change of ventilator circuit no more often than every 48 hours, and performance of hand hygiene before and after contact with each patient.

## ACUTE RESPIRATORY FAILURE IN COPD

In patients with essentially normal lung tissue, ARF usually means  $\text{Paco}_2$  above 50 mm Hg and  $\text{Pao}_2$  below 50 mm Hg. These limits, however, don't apply to patients with COPD, who usually have a consistently high  $\text{Paco}_2$  and low  $\text{Pao}_2$ . In patients with COPD, only acute deterioration in ABG values, with corresponding clinical deterioration, indicates ARF.

## **Causes and Incidence**

ARF may develop in patients with COPD as a result of any condition that increases the work of breathing and decreases the respiratory drive. Such conditions include respiratory tract infection (such as bronchitis or pneumonia). The most common precipitating factor is bronchospasm, or accumulating secretions secondary to cough suppression. Other causes of ARF in COPD include the following:

- ◆ CNS depression—head trauma or injudicious use of sedatives, opioids, tranquilizers, or oxygen ( $\text{O}_2$ )
- ◆ Cardiovascular disorders—myocardial infarction, heart failure, or pulmonary emboli
- ◆ Airway irritants—smoke or fumes
- ◆ Endocrine and metabolic disorders—myxedema or metabolic alkalosis
- ◆ Thoracic abnormalities—chest trauma, pneumothorax, or thoracic or abdominal surgery

The incidence of ARF increases markedly with age and is especially high among people age 65 and older.

## **Pathophysiology**

An acute and progressive exacerbation of COPD is triggered by the cessation of maintenance medications or some type of infection. Damage to the epithelium from ongoing exposure to noxious gases or particles impairs the mucociliary response causing the accumulation of mucus and bacteria and contributing to the obstruction of airways. The permeant enlargement of the airspaces by the terminal bronchioles leads to a decrease in the alveolar surface area for gas exchange and contributes to ineffective ventilation.

## **Complications**

- ◆ Respiratory failure
- ◆ Pneumonia

- ◆ Hypoxemia
- ◆ Pneumothorax
- ◆ Heart failure

## Signs And Symptoms

In patients who have COPD with ARF, increased ventilation–perfusion mismatch and reduced alveolar ventilation decrease  $\text{Pao}_2$  (hypoxemia) and increase  $\text{Paco}_2$  (hypercapnia). This rise in  $\text{CO}_2$  lowers the pH. The resulting hypoxemia and acidemia affect all body organs, especially the CNS and the respiratory and cardiovascular systems.

Specific symptoms vary with the underlying cause of ARF but may include these systems:

- ◆ Respiratory—Rate may be increased, decreased, or normal depending on the cause; respirations may be shallow, deep, or alternate between the two; and air hunger may occur. Cyanosis may or may not be present, depending on the Hb level and arterial oxygenation. Auscultation of the chest may reveal crackles, rhonchi, wheezing, or diminished breath sounds.
- ◆ CNS—When hypoxemia and hypercapnia occur, the patient may show evidence of restlessness, confusion, loss of concentration, irritability, tremulousness, diminished tendon reflexes, and papilledema; the patient may slip into a coma.
- ◆ Cardiovascular—Tachycardia, with increased cardiac output and mildly elevated blood pressure secondary to adrenal release of catecholamine, occurs early in response to low  $\text{Pao}_2$ . With myocardial hypoxia, arrhythmias may develop. Pulmonary hypertension, secondary to pulmonary capillary vasoconstriction, may cause increased pressures on the right side of the heart, jugular vein distention, an enlarged liver, and peripheral edema. Stresses on the heart may precipitate cardiac failure.

## Diagnosis

Progressive deterioration in ABG levels and pH, when compared with the patient’s “normal” values, strongly suggests ARF in COPD. (In patients with essentially normal lung tissue, pH below 7.35 usually indicates ARF, but patients with COPD display an even greater deviation from this normal value, as they do with  $\text{Paco}_2$  and  $\text{Pao}_2$ .)

Other supporting findings include:

- ◆ Bicarbonate—Increased levels indicate metabolic alkalosis or reflect metabolic compensation for chronic respiratory acidosis.
- ◆ Hematocrit (HCT) and Hb—Abnormally low levels may be due to blood loss, indicating decreased oxygen-carrying capacity. Elevated levels may occur with chronic hypoxemia.
- ◆ Serum electrolytes—Hypokalemia and hypochloremia may result from diuretic and corticosteroid therapies used to treat ARF.
- ◆ White blood cell count—Count is elevated if ARF is due to bacterial infection; Gram stain and sputum culture can identify pathogens.
- ◆ Chest X-ray—Findings identify pulmonary pathologic conditions, such as emphysema, atelectasis, lesions, pneumothorax, infiltrates, or effusions.
- ◆ Electrocardiogram—Arrhythmias commonly suggest cor pulmonale and myocardial hypoxia.

## Treatment

ARF in patients with COPD is an emergency that requires cautious O<sub>2</sub> therapy (using nasal prongs or Venturi mask) to raise the Pao<sub>2</sub>. In patients with chronic hypercapnia, O<sub>2</sub> therapy can cause hypoventilation by increasing Paco<sub>2</sub> and decreasing the respiratory drive, necessitating mechanical ventilation. The minimum fraction of inspired air (FIO<sub>2</sub>) required to maintain ventilation or O<sub>2</sub> saturation greater than 85% to 90% should be used. If significant uncompensated respiratory acidosis or unrefractory hypoxemia exists, mechanical ventilation (through an ET or a tracheostomy tube) or noninvasive ventilation (with a face or nose mask) may be necessary. Treatment routinely includes antibiotics for infection, bronchodilators, and possibly steroids.

## Special Considerations

- ◆ Because most patients with ARF are treated in an intensive care unit, orient them to the environment, procedures, and routines to minimize their anxiety.
- ◆ To reverse hypoxemia, administer O<sub>2</sub> at appropriate concentrations to maintain Pao<sub>2</sub> at a minimum of 50 to 60 mm Hg. Patients with COPD usually require only small amounts of supplemental O<sub>2</sub>. Watch for a positive response—such as improvement in the patient's breathing, color, and ABG levels.

- ◆ Maintain a patent airway. If the patient is retaining CO<sub>2</sub>, encourage them to cough and to breathe deeply. Teach them to use pursed-lip and diaphragmatic breathing to control dyspnea. If the patient is alert, have them use an incentive spirometer; if they are intubated and lethargic, turn the patient every 1 to 2 hours. Use postural drainage and chest physiotherapy to help clear secretions.
- ◆ In an intubated patient, suction the trachea as needed after hyperoxygenation. Observe for a change in quantity, consistency, and color of sputum. Provide humidification to liquefy secretions.
- ◆ Observe the patient closely for respiratory arrest. Auscultate for chest sounds. Monitor ABG levels and report any changes immediately.
- ◆ Check the cardiac monitor for arrhythmias.

If the patient requires mechanical ventilation:

- ◆ Check ventilator settings, cuff pressures, and ABG values often because the FIO<sub>2</sub> setting depends on ABG levels. Draw specimens for ABG analysis 20 to 30 minutes after every FIO<sub>2</sub> change or oximetry check.
- ◆ Prevent infection by performing hand hygiene and using sterile technique while suctioning.
- ◆ Stress ulcers are common in the intubated patient. Check gastric secretions for evidence of bleeding if the patient has a nasogastric (NG) tube or if the patient complains of epigastric tenderness, nausea, or vomiting. Monitor Hb level and HCT; check all stools for occult blood. Administer antacids, histamine-2 receptor antagonists, or sucralfate, as ordered.
- ◆ To prevent nasal necrosis, keep the nasotracheal tube midline within the patient's nostrils and provide good hygiene. Loosen the tape periodically to prevent skin breakdown. Avoid excessive movement of any tubes; make sure the ventilator tubing is adequately supported.



## PREVENTION

- ◆ *To prevent VAP, implement Ventilator Bundle.*
- ◆ *Prevent tracheal erosion, which can result from artificial airway cuff overinflation. Use the minimal leak technique and a cuffed tube with high residual volume (low-pressure cuff), a foam cuff, or a pressure-regulating valve on the cuff.*

- To prevent oral or vocal cord trauma, make sure that the ET tube is positioned midline or moved carefully from side to side every 8 hours.

## PULMONARY EDEMA

Pulmonary edema is the accumulation of fluid in the extravascular spaces of the lung. In cardiogenic pulmonary edema, fluid accumulation results from elevations in pulmonary venous and capillary hydrostatic pressures. A common complication of cardiac disorders, pulmonary edema can occur as a chronic condition or it can develop quickly to cause death. (See *How pulmonary edema develops*, page 131.)

### Causes And Incidence

Pulmonary edema usually results from left-sided heart failure due to arteriosclerotic, hypertensive, cardiomyopathic, or valvular cardiac disease. In such disorders, the compromised left ventricle is unable to maintain adequate cardiac output; increased pressures are transmitted to the left atrium, pulmonary veins, and pulmonary capillary bed. This increased pulmonary capillary hydrostatic force promotes transudation of intravascular fluids into the pulmonary interstitium, decreasing lung compliance and interfering with gas exchange. Other factors that may predispose the patient to pulmonary edema include:

- ◆ excessive infusion of I.V. fluids
- ◆ decreased serum colloid osmotic pressure as a result of nephrosis, protein-losing enteropathy, extensive burns, hepatic disease, or nutritional deficiency
- ◆ impaired lung lymphatic drainage from Hodgkin lymphoma or obliterative lymphangitis after radiation
- ◆ mitral stenosis, which impairs left atrial emptying
- ◆ pulmonary veno-occlusive disease
- ◆ lung damage from a severe infection or exposure to poisonous gas
- ◆ kidney failure

### Pathophysiology

A hemodynamic disturbance or alteration in the permeability of the microvasculature allowing fluid to pass through into the interstitial space. This interstitial edema progresses when the capacity of the lymphatics is

exceeded and unable to drain the fluid efficiently and subsequently decreasing lung compliance and shortness of breath.

## Complications

- ◆ respiratory failure
- ◆ pleural effusion
- ◆ edema to lower extremities and abdomen
- ◆ death

## Signs and Symptoms

The early symptoms of pulmonary edema reflect interstitial fluid accumulation and diminished lung compliance: dyspnea on exertion, paroxysmal nocturnal dyspnea, orthopnea, and coughing. Clinical features include tachycardia, tachypnea, dependent crackles, jugular vein distention, and a diastolic ( $S_3$ ) gallop. With severe pulmonary edema, the alveoli and bronchioles may fill with fluid and intensify the early symptoms. Respiration becomes labored and rapid, with more diffuse crackles and coughing that produces frothy, bloody sputum. Tachycardia increases, and arrhythmias may occur. Skin becomes cold, clammy, diaphoretic, and cyanotic. Blood pressure falls and the pulse becomes thready as cardiac output falls.

Symptoms of severe heart failure with pulmonary edema may also include signs of hypoxemia, such as anxiety, restlessness, and changes in the patient's level of consciousness.

## Diagnosis

Clinical features of pulmonary edema permit a working diagnosis. ABG analysis usually shows hypoxia; the  $Paco_2$  is variable. Profound respiratory alkalosis and acidosis may occur. Chest X-ray shows diffuse haziness of the lung fields and, commonly, cardiomegaly and pleural effusions. Ultrasound (echocardiogram) may show weak heart muscle, leaking or narrow heart valves, and fluid surrounding the heart. Pulmonary artery catheterization helps identify left-sided heart failure by showing elevated PAWPs. This helps to rule out ARDS—in which pulmonary wedge pressure is usually normal.

## Treatment

Treatment measures for pulmonary edema are designed to reduce extravascular fluid, improve gas exchange and myocardial function and, if

possible, correct any underlying pathologic conditions.

Administration of high concentrations of oxygen by a cannula, a face mask and, if the patient fails to maintain an acceptable  $\text{PaO}_2$  level, assisted ventilation improves oxygen delivery to the tissues and usually improves acid-base disturbances. Diuretics—furosemide and bumetanide, for example—promote diuresis, which reduces extravascular fluid.

Treatment of heart failure includes angiotensin-converting enzyme inhibitors, diuretics, inotropic drugs such as digoxin, antiarrhythmic agents, beta-adrenergic blockers, and human B-type natriuretic peptide. Vasodilator drugs, such as nitroprusside, may be used to reduce preload and afterload in acute episodes of pulmonary edema.

Morphine is used to reduce anxiety and dyspnea as well as dilate the systemic venous bed, promoting blood flow from pulmonary circulation to the periphery.

## **Special Considerations**

- ◆ Carefully monitor the vulnerable patient for early signs of pulmonary edema, especially tachypnea, tachycardia, and abnormal breath sounds. Report any abnormalities. Assess for peripheral edema and weight gain, which may also indicate that fluid is accumulating in tissue.
- ◆ Administer oxygen as ordered.
- ◆ Monitor the patient's vital signs every 15 to 30 minutes while administering nitroprusside in dextrose 5% in water by I.V. drip. Protect the nitroprusside solution from light by wrapping the bottle or bag with aluminum foil and discard unused solution after 4 hours. Watch for arrhythmias in the patient receiving cardiac glycosides and for marked respiratory depression in the patient receiving morphine.
- ◆ Assess the patient's condition frequently, and record response to treatment. Monitor ABG levels, oral and I.V. fluid intake, urine output and, in the patient with a pulmonary artery catheter, pulmonary end-diastolic and wedge pressures. Check the cardiac monitor often. Report changes immediately.
- ◆ Carefully record the time and amount of morphine given.
- ◆ Reassure the patient, who will be anxious because of hypoxia and respiratory distress. Explain all procedures. Provide emotional support to the family as well.

## **COR PULMONALE**

The World Health Organization defines chronic cor pulmonale as “hypertrophy of the right ventricle resulting from diseases affecting the function or the structure of the lungs, except when these pulmonary alterations are the result of diseases that primarily affect the left side of the heart or of congenital heart disease.” Invariably, cor pulmonale follows some disorder of the lungs, pulmonary vessels, chest wall, or respiratory control center. For instance, COPD produces pulmonary hypertension, which leads to right ventricular hypertrophy and right-sided heart failure. Because cor pulmonale generally occurs late during the course of COPD and other irreversible diseases, the prognosis is generally poor.

## **Causes and Incidence**

Approximately 85% of patients with cor pulmonale have COPD, and 25% of patients with COPD eventually develop cor pulmonale.

Other respiratory disorders that produce cor pulmonale include:

- ◆ obstructive lung diseases—for example, bronchiectasis and cystic fibrosis
- ◆ restrictive lung diseases—for example, pneumoconiosis, interstitial pneumonitis, scleroderma, and sarcoidosis
- ◆ loss of lung tissue after extensive lung surgery
- ◆ congenital cardiac shunts—such as a ventricular septal defect
- ◆ pulmonary vascular diseases—for example, recurrent thromboembolism, primary pulmonary hypertension, schistosomiasis, and pulmonary vasculitis
- ◆ respiratory insufficiency without pulmonary disease—for example, in chest wall disorders such as kyphoscoliosis, neuromuscular incompetence due to muscular dystrophy and amyotrophic lateral sclerosis, polymyositis, and spinal cord lesions above C6
- ◆ obesity hypoventilation syndrome (pickwickian syndrome) and upper airway obstruction
- ◆ living at high altitudes (chronic mountain sickness)

Cor pulmonale accounts for about 25% of all types of heart failure. It's most common in areas of the world where the incidence of cigarette smoking and COPD is high; cor pulmonale affects middle-aged to elderly men more often than women, but the incidence in women is increasing. In children, cor pulmonale may be a complication of cystic fibrosis, hemosiderosis, upper airway obstruction, scleroderma, extensive bronchiectasis, neurologic

diseases affecting respiratory muscles, or abnormalities of the respiratory control center.

## **Pathophysiology**

Pulmonary capillary destruction and pulmonary vasoconstriction (usually secondary to hypoxia) reduce the area of the pulmonary vascular bed. Thus, pulmonary vascular resistance is increased, causing pulmonary hypertension. To compensate for the extra work needed to force blood through the lungs, the right ventricle dilates and hypertrophies. In response to low oxygen content, the bone marrow produces more red blood cells (RBCs), causing erythrocytosis. When the HCT exceeds 55%, blood viscosity increases, which further aggravates pulmonary hypertension and increases the hemodynamic load on the right ventricle. Right-sided heart failure is the result.

## **Complications**

- ◆ Right- and left-sided heart failure
- ◆ Hepatomegaly
- ◆ Edema
- ◆ Ascites
- ◆ Pleural effusions
- ◆ Thromboembolism

## **Signs and Symptoms**

As long as the heart can compensate for the increased pulmonary vascular resistance, clinical features reflect the underlying disorder and occur mostly in the respiratory system. They include chronic productive cough, exertional dyspnea, wheezing respirations, fatigue, and weakness. Progression of cor pulmonale is associated with dyspnea (even at rest) that worsens on exertion, tachypnea, orthopnea, edema, weakness, and right upper quadrant discomfort. Chest examination reveals findings characteristic of the underlying lung disease.

Signs of cor pulmonale and right-sided heart failure include dependent edema; distended jugular veins; prominent parasternal or epigastric cardiac impulse; hepatojugular reflux; an enlarged, tender liver; ascites; and tachycardia. Decreased cardiac output may cause a weak pulse and hypotension. Chest examination yields various findings, depending on the underlying cause of cor pulmonale.

In COPD, auscultation reveals wheezing, rhonchi, and diminished breath sounds. When the disease is secondary to upper airway obstruction or damage to CNS respiratory centers, chest findings may be normal, except for a right ventricular lift, gallop rhythm, and loud pulmonic component of S<sub>2</sub>. Tricuspid insufficiency produces a pansystolic murmur heard at the lower left sternal border; its intensity increases on inspiration, distinguishing it from a murmur due to mitral valve disease. A right ventricular early murmur that increases on inspiration can be heard at the left sternal border or over the epigastrium. A systolic pulmonic ejection click may also be heard. Alterations in the patient's level of consciousness may occur.

## Diagnosis

- ◆ PAP measurements show increased right ventricular and PAPs, stemming from increased pulmonary vascular resistance. Right ventricular systolic and pulmonary artery systolic pressures will exceed 30 mm Hg. Pulmonary artery diastolic pressure will exceed 15 mm Hg.
- ◆ Echocardiography or angiography indicates right ventricular enlargement; echocardiography can estimate PAP while also ruling out structural and congenital lesions.
- ◆ Chest X-ray shows large central pulmonary arteries and suggests right ventricular enlargement by rightward enlargement of the heart's silhouette on an anterior chest film.
- ◆ ABG analysis shows decreased Pao<sub>2</sub> (typically less than 70 mm Hg and usually no more than 90 mm Hg on room air).
- ◆ Electrocardiogram frequently shows arrhythmias, such as premature atrial and ventricular contractions and atrial fibrillation during severe hypoxia; it may also show right bundle-branch block, right axis deviation, prominent P waves and inverted T wave in right precordial leads, and right ventricular hypertrophy.
- ◆ PFTs show results consistent with the underlying pulmonary disease.
- ◆ HCT is typically greater than 50%.

## Treatment

Treatment of cor pulmonale is designed to reduce hypoxemia, increase the patient's exercise tolerance and, when possible, correct the underlying condition.

In addition to bed rest, treatment may include administration of:

- ◆ a cardiac glycoside (digoxin)
- ◆ antibiotics when respiratory infection is present; culture and sensitivity of a sputum specimen helps select an antibiotic
- ◆ potent pulmonary artery vasodilators (such as diazoxide, nitroprusside, hydralazine, angiotensin-converting enzyme inhibitors, calcium channel blockers, or prostaglandins) in primary pulmonary hypertension
- ◆ oxygen by mask or cannula in concentrations ranging from 24% to 40%, depending on  $\text{Pao}_2$ , as necessary; in acute cases, therapy may also include mechanical ventilation; patients with underlying COPD generally shouldn't receive high concentrations of oxygen because of possible subsequent respiratory depression
- ◆ a low-sodium diet, restricted fluid intake, and diuretics, such as furosemide, to reduce edema
- ◆ phlebotomy to reduce the RBC count
- ◆ anticoagulants to reduce the risk of thromboembolism

Depending on the underlying cause, some variations in treatment may be indicated. For example, a tracheotomy may be necessary if the patient has an upper airway obstruction. Steroids may be used in the patient with a vasculitis autoimmune phenomenon or acute exacerbations of COPD.

## **Special Considerations**

- ◆ Plan diet carefully with the patient and staff dietitian. Because the patient may lack energy and tire easily when eating, provide small, frequent feedings rather than three heavy meals.
- ◆ Prevent fluid retention by limiting the patient's fluid intake to 1 to 2 qt (1 to 2 L)/day and providing a low-sodium diet.
- ◆ Monitor serum potassium levels closely if the patient is receiving diuretics. Low serum potassium levels can increase the risk of arrhythmias associated with cardiac glycosides.
- ◆ Watch the patient for signs of digoxin toxicity, such as complaints of anorexia, nausea, vomiting, and halos around visual images and color perception shifts. Monitor for cardiac arrhythmias. Teach the patient to check their radial pulse before taking digoxin or any cardiac glycoside. They should be instructed to notify the physician if they detect changes in pulse rate.
- ◆ Reposition bedridden patients often to prevent atelectasis.

- ◆ Provide meticulous respiratory care, including oxygen therapy and, for the patient with COPD, pursed-lip breathing exercises. Periodically measure ABG levels and watch for signs of respiratory failure: changes in pulse rate, labored respirations, changes in mental status, and increased fatigue after exertion.

Before discharge, maintain the following protocol:

- ◆ Make sure that the patient understands the importance of maintaining a low-sodium diet, weighing himself daily, and watching for increased edema. Teach patient to detect edema by pressing the skin over a shin with one finger, holding it for a second or two, then checking for a finger impression. Increased weight, increased edema, or respiratory difficulty should be reported to the healthcare provider.
- ◆ Instruct the patient to plan for frequent rest periods and to do breathing exercises regularly.
- ◆ If the patient needs supplemental oxygen therapy at home, refer them to an agency that can help obtain the required equipment and, as necessary, arrange for follow-up examinations.
- ◆ If the patient has been placed on anticoagulant therapy, emphasize the need to watch for bleeding (epistaxis, hematuria, bruising) and to report signs to the physician. Also encourage patient to return for periodic laboratory tests to monitor partial thromboplastin time (PTT), fibrinogen level, platelet count, HCT, Hb level, and prothrombin time.
- ◆ Because pulmonary infection commonly exacerbates COPD and cor pulmonale, tell the patient to watch for and immediately report early signs of infection, such as increased sputum production, change in sputum color, increased coughing or wheezing, chest pain, fever, and tightness in the chest. Tell the patient to avoid crowds and persons known to have pulmonary infections, especially during the flu season. The patient should receive pneumovax and annual influenza vaccines.
- ◆ Warn the patient to avoid substances that may depress the ventilatory drive, such as sedatives and alcohol.

## **LEGIONNAIRES' DISEASE**

Legionnaires' disease is an acute bronchopneumonia produced by a gram-negative bacillus, *Legionella pneumophila*. It derives its name and notoriety from the peculiar, highly publicized disease that struck 182 people (29 of whom died) at an American Legion convention in Philadelphia in July 1976.

This disease may occur epidemically or sporadically, usually in late summer or early fall. Its severity ranges from a mild illness, with or without pneumonitis, to multilobar pneumonia, with a mortality as high as 15%. A milder, self-limiting form (Pontiac syndrome) subsides within a few days but leaves the patient fatigued for several weeks. This form mimics Legionnaires' disease but produces few or no respiratory symptoms, no pneumonia, and no fatalities.

## Causes and Incidence

*Legionella pneumophila* is an aerobic, gram-negative bacillus that's probably transmitted by an airborne route. In past epidemics, it has spread through cooling towers or evaporation condensers in air-conditioning systems. However, *Legionella* bacilli also flourish in soil and excavation sites. The disease doesn't spread from person to person.

Legionnaires' disease is most likely to affect:

- ◆ middle-aged and elderly people
- ◆ immunocompromised patients (particularly those receiving corticosteroids, e.g., after a transplant) or those with lymphoma or other disorders associated with delayed hypersensitivity
- ◆ patients with a chronic underlying disease, such as diabetes, chronic renal failure, or COPD
- ◆ those with alcoholism
- ◆ cigarette smokers
- ◆ those on a ventilator for extended periods

## Pathophysiology

When water droplets containing a sufficient amount of the *Legionella* bacterium enter the atmosphere they can be inhaled into the lungs. There they invade the epithelial cells of the lungs and begin to replicate intracellularly causing a Legionnaires' infection.

## Complications

- ◆ Respiratory failure
- ◆ Septic shock
- ◆ Acute kidney failure

## Signs and Symptoms

The multisystem clinical features of Legionnaires' disease follow a predictable sequence, although the onset of the disease may be gradual or sudden. After a 2- to 10-day incubation period, nonspecific, prodromal signs and symptoms appear, including diarrhea, anorexia, malaise, diffuse myalgias and generalized weakness, headache, and recurrent chills. An unremitting fever develops within 12 to 48 hours with a temperature that may reach 105° F (40.6° C). A cough then develops that's nonproductive initially but eventually may produce grayish, nonpurulent, and occasionally blood-streaked sputum.

Other characteristic features include nausea, vomiting, disorientation, mental sluggishness, confusion, mild temporary amnesia, pleuritic chest pain, tachypnea, dyspnea, and fine crackles. Patients who develop pneumonia may also experience hypoxia. Other complications include hypotension, delirium, heart failure, arrhythmias, ARF, renal failure, and shock (usually fatal).

## Diagnosis

The patient history focuses on possible sources of infection and predisposing conditions. Additional tests reveal the following:

- ◆ Chest X-ray shows patchy, localized infiltration, which progresses to multilobar consolidation (usually involving the lower lobes), pleural effusion and, in fulminant disease, opacification of the entire lung.
- ◆ Auscultation reveals fine crackles, progressing to coarse crackles as the disease advances.
- ◆ Abnormal findings include leukocytosis, increased erythrocyte sedimentation rate, an increase in liver enzyme levels (alanine aminotransferase, aspartate aminotransferase, and alkaline phosphatase), hyponatremia, decreased Pao<sub>2</sub> and, initially, decreased Paco<sub>2</sub>. Bronchial washings and blood, pleural fluid, and sputum tests rule out other infections.

**Dx CONFIRMING DIAGNOSIS** *Definitive tests include direct immunofluorescence of respiratory tract secretions and tissue, culture of L. pneumophila, and indirect fluorescent antibody testing of serum comparing acute samples with convalescent samples drawn at least 3 weeks later. A convalescent serum showing a fourfold or greater rise in antibody titer for Legionella confirms the diagnosis.*

## Treatment

Antibiotic treatment begins as soon as Legionnaires' disease is suspected and diagnostic material is collected; it shouldn't await laboratory confirmation. A quinolone (ciprofloxacin, levofloxacin, moxifloxacin, or gatifloxacin) is commonly used, although a macrolide (azithromycin, clarithromycin, or erythromycin) may be prescribed for some patients. Supportive therapy includes administration of antipyretics, fluid replacement, circulatory support with pressor drugs, if necessary, and oxygen administration by mask, cannula, or mechanical ventilation.

## **Special Considerations**

- ◆ Closely monitor the patient's respiratory status. Evaluate chest wall expansion, depth and pattern of respirations, cough, and chest pain. Watch for restlessness as a sign of hypoxemia, which requires suctioning, repositioning, or more aggressive oxygen therapy.
- ◆ Continually monitor the patient's vital signs, oximetry or ABG values, level of consciousness, and dryness and color of lips and mucous membranes. Watch for signs of shock (decreased blood pressure, thready pulse, diaphoresis, and clammy skin).
- ◆ Keep the patient comfortable. Provide mouth care frequently. If necessary, apply soothing cream to the nostrils.
- ◆ Replace fluid and electrolytes, as needed. The patient with renal failure may require dialysis.
- ◆ Provide mechanical ventilation and other respiratory therapy, as needed. Teach the patient how to cough effectively and encourage deep-breathing exercises. Stress the need to continue these until recovery is complete.
- ◆ Give antibiotic therapy as indicated and observe carefully for adverse effects.

## **ATELECTASIS**

Atelectasis is incomplete expansion of lobules (clusters of alveoli) or lung segments, which may result in partial or complete lung collapse. Because parts of the lung are unavailable for gas exchange, unoxygenated blood passes through these areas unchanged, resulting in hypoxemia. Atelectasis may be chronic or acute. Many patients undergoing upper abdominal or thoracic surgery experience atelectasis to some degree. The prognosis depends on prompt removal of any airway obstruction, relief of hypoxemia, and re-expansion of the collapsed lung.

## Causes

Atelectasis commonly results from bronchial occlusion by mucus plugs. It's a problem in many patients with COPD, bronchiectasis, or cystic fibrosis and in those who smoke heavily. (Smoking increases mucus production and damages cilia.) Atelectasis may also result from occlusion by foreign bodies, bronchogenic carcinoma, and inflammatory lung disease.

Other causes include RDS of the neonate (hyaline membrane disease), oxygen toxicity, and pulmonary edema, in which alveolar surfactant changes increase surface tension and permit complete alveolar deflation.

External compression, which inhibits full lung expansion, or any condition that makes deep breathing painful may also cause atelectasis. Such compression or pain may result from abdominal surgical incisions, rib fractures, pleuritic chest pain, tight dressings around the chest, stab wounds, impalement accidents, car accidents in which the driver slams into the steering column, or obesity (which elevates the diaphragm and reduces tidal volume).

Prolonged immobility may also cause atelectasis by producing preferential ventilation of one area of the lung over another. Mechanical ventilation using constant small tidal volumes without intermittent deep breaths may also result in atelectasis. CNS depression (as in drug overdose) eliminates periodic sighing and is a predisposing factor of progressive atelectasis.

## Pathophysiology

Atelectasis results from some type of obstruction or compression of the lungs or bronchus. Retraction of the lung occurs when the blood circulating in the alveolar capillary bed absorbs the gas from the alveolar in the unventilated lung. The alveolar spaces fill with secretions and cells, preventing the complete collapse of the lung. The surrounding tissues distend and displace, shifting the heart and mediastinum toward the atelectatic area.

## Complications

- ◆ Respiratory failure
- ◆ Pneumonia
- ◆ Hypoxemia

## Signs and Symptoms

Clinical effects vary with the cause of collapse, the degree of hypoxemia, and any underlying disease but generally include some degree of dyspnea. Atelectasis of a small area of the lung may produce only minimal symptoms that subside without specific treatment. However, massive collapse can produce severe dyspnea, anxiety, cyanosis, diaphoresis, peripheral circulatory collapse, tachycardia, and substernal or intercostal retraction. Also, atelectasis may result in compensatory hyperinflation of unaffected areas of the lung, mediastinal shift to the affected side, and elevation of the ipsilateral hemidiaphragm.

## Diagnosis

Diagnosis requires an accurate patient history, a physical examination, and a chest X-ray. Auscultation reveals diminished or bronchial breath sounds. When much of the lung is collapsed, percussion reveals dullness. However, extensive areas of “microatelectasis” may exist without abnormalities on the chest X-ray. In widespread atelectasis, the chest X-ray shows characteristic horizontal lines in the lower lung zones. With segmental or lobar collapse, characteristic dense shadows commonly associated with hyperinflation of neighboring lung zones are also apparent. If the cause is unknown, diagnostic procedures may include bronchoscopy to rule out an obstructing neoplasm or a foreign body.

## Treatment

Treatment includes incentive spirometry, frequent coughing, and deep-breathing exercises. If atelectasis is secondary to mucus plugging, mucolytics, chest percussion, and postural drainage may be used. If these measures fail, bronchoscopy may be helpful in removing secretions. Humidity and bronchodilators can improve mucociliary clearance and dilate airways.

Atelectasis secondary to an obstructing neoplasm may require surgery or radiation therapy. Postoperative thoracic and abdominal surgery patients require analgesics to facilitate deep breathing, which minimizes the risk of atelectasis.

## Special Considerations

- ♦ If mechanical ventilation is used, tidal volume should be maintained at appropriate levels to ensure adequate expansion of the lungs. Use the sigh mechanism on the ventilator, if appropriate, to intermittently increase tidal

volume at the rate of 10 to 15 sighs/hour. Implement the Ventilator Bundle to prevent VAP.

- ◆ Use an incentive spirometer to encourage deep inspiration through positive reinforcement. Teach the patient how to use the spirometer and encourage them to use it every 1 to 2 hours.
- ◆ Humidify inspired air and encourage adequate fluid intake to mobilize secretions. To promote loosening and clearance of secretions, encourage deep-breathing and coughing exercises and use postural drainage and chest percussion.
- ◆ If the patient is intubated or uncooperative, provide suctioning, as needed. Use sedatives with discretion because they depress respirations and the cough reflex as well as suppress sighing. However, remember that the patient won't cooperate with treatment if they are in pain.
- ◆ Assess breath sounds and ventilatory status frequently; report changes at once.
- ◆ Teach the patient about respiratory care, including postural drainage, coughing, and deep breathing.
- ◆ Encourage the patient to stop smoking and lose weight, as needed. Refer them to appropriate support groups for help.
- ◆ Provide reassurance and emotional support; the patient may be anxious because of hypoxia or respiratory distress.



### PREVENTION

- ◆ *In a patient who is bedridden, encourage movement and deep breathing.*
- ◆ *Administer adequate analgesics.*
- ◆ *To prevent atelectasis, encourage the postoperative or other high-risk patient to cough and deep-breathe every 1 to 2 hours. To minimize pain during coughing exercises, splint the incision; teach the patient this technique as well. Gently reposition the patient often and encourage ambulation as soon as possible.*
- ◆ *Teach patients to keep small objects out of reach of children.*

## RESPIRATORY ACIDOSIS

An acid–base disturbance characterized by reduced alveolar ventilation and manifested by hypercapnia ( $\text{Paco}_2$  greater than 45 mm Hg), respiratory acidosis can be acute (because of a sudden failure in ventilation) or chronic

(as in long-term pulmonary disease). The prognosis depends on the severity of the underlying disturbance as well as the patient's general clinical condition.

## Causes and Incidence

Some predisposing factors in respiratory acidosis include:

- ◆ Drugs—Opioids, anesthetics, hypnotics, and sedatives, including some of the new designer drugs, such as Ecstasy, decrease the sensitivity of the respiratory center.
- ◆ CNS trauma—Medullary injury may impair ventilatory drive.
- ◆ Chronic metabolic alkalosis—Respiratory compensatory mechanisms attempt to normalize pH by decreasing alveolar ventilation.
- ◆ Ventilation therapy—Use of high-flow oxygen ( $O_2$ ) in chronic respiratory disorders suppresses the patient's hypoxic drive to breathe.
- ◆ Neuromuscular diseases (such as myasthenia gravis, Guillain–Barré syndrome, and poliomyelitis)—Failure of the respiratory muscles to respond properly to respiratory drive decreases alveolar ventilation.
- ◆ In addition, respiratory acidosis can result from airway obstruction or parenchymal lung disease, which interferes with alveolar ventilation; COPD; asthma; severe acute respiratory distress syndrome (SARS); chronic bronchitis; large pneumothorax; extensive pneumonia; and pulmonary edema.

Hypoventilation compromises elimination of  $CO_2$  produced through metabolism. The retained  $CO_2$  then combines with water to form an excess of carbonic acid, decreasing the blood pH. As a result, the concentration of hydrogen ions in body fluids, which directly reflects acidity, increases.

## Pathophysiology

Lung diseases or conditions that cause hypoventilation result in  $CO_2$  being produced at a rapid rate. Lack of adequate ventilation quickly increases the partial pressure of arterial  $CO_2$ . The increase in  $Paco_2$  also results in the decrease in bicarbonate ration and decreasing the pH to an acidotic state.

## Complications

- ◆ Shock
- ◆ Cardiac arrest

## Signs and Symptoms

Acute respiratory acidosis produces CNS disturbances that reflect changes in the pH of cerebrospinal fluid rather than increased CO<sub>2</sub> levels in cerebral circulation. Effects range from restlessness, confusion, and apprehension to somnolence, with a fine or flapping tremor (asterixis), or coma. The patient may complain of headaches as well as exhibiting dyspnea and tachypnea with papilledema and depressed reflexes. Unless the patient is receiving O<sub>2</sub>, hypoxemia accompanies respiratory acidosis. This disorder may also cause cardiovascular abnormalities, such as tachycardia, hypertension, atrial and ventricular arrhythmias and, in severe acidosis, hypotension with vasodilation (bounding pulses and warm periphery).

## Diagnosis

 **CONFIRMING DIAGNOSIS** ABG analysis confirms the diagnosis: Paco<sub>2</sub> exceeds the normal 45 mm Hg; pH is below the normal range of 7.35 to 7.45 unless compensation has occurred; and bicarbonate is normal in the acute stage but elevated in the chronic stage.

Chest X-ray, CT scan, and PFTs can help determine the cause.

## Treatment

Effective treatment of respiratory acidosis requires correction of the underlying source of alveolar hypoventilation.

Significantly reduced alveolar ventilation may require mechanical ventilation until the underlying condition can be treated. In COPD, this includes bronchodilators, O<sub>2</sub>, corticosteroids, and antibiotics for infectious conditions; drug therapy for conditions such as myasthenia gravis; removal of foreign bodies from the airway; antibiotics for pneumonia; dialysis or charcoal to remove toxic drugs; and correction of metabolic alkalosis.

Dangerously low blood pH (less than 7.15) can produce profound CNS and cardiovascular deterioration; careful administration of I.V. sodium bicarbonate may be required. In chronic lung disease, elevated CO<sub>2</sub> may persist despite optimal treatment.

## Special Considerations

- ◆ Be alert for critical changes in the patient's respiratory, CNS, and cardiovascular functions. Report such changes as well as any variations in ABG values or electrolyte status immediately. Also, maintain adequate hydration.
- ◆ Maintain a patent airway and provide adequate humidification if acidosis requires mechanical ventilation. Perform tracheal suctioning regularly and vigorous chest physiotherapy if ordered. Continuously monitor ventilator settings and respiratory status.
- ◆ To prevent respiratory acidosis, closely monitor patients with COPD and chronic CO<sub>2</sub> retention for signs of acidosis. Also, administer O<sub>2</sub> at low flow rates; closely monitor all patients who receive opioids and sedatives. Instruct patients who have received general anesthesia to turn, cough, and perform deep-breathing exercises frequently to prevent the onset of respiratory acidosis.

## **RESPIRATORY ALKALOSIS**

Respiratory alkalosis is an acid–base disturbance characterized by a decrease in the Paco<sub>2</sub> to less than 35 mm Hg, which is due to alveolar hyperventilation. Uncomplicated respiratory alkalosis leads to a decrease in hydrogen ion concentration, which results in elevated blood pH. Hypocapnia occurs when the elimination of CO<sub>2</sub> by the lungs exceeds the production of CO<sub>2</sub> at the cellular level.

### **Causes**

Causes of respiratory alkalosis fall into two categories:

- ◆ pulmonary—severe hypoxemia, pneumonia, interstitial lung disease, pulmonary vascular disease, and acute asthma
- ◆ nonpulmonary—anxiety, fever, aspirin toxicity, metabolic acidosis, CNS disease (inflammation or tumor), sepsis, hepatic failure, and pregnancy

### **Pathophysiology**

An underlying condition or stimulus that causes hyperventilation, expels an increased amount of CO<sub>2</sub>. CO<sub>2</sub> in the circulation is shifted causing hydrogen ions and bicarbonate to change into additional CO<sub>2</sub> via the enzyme carbonic anhydrase which, in turn, decreases the available hydrogen ions and increasing the pH.

## Complications

- ◆ Cardiac arrhythmias
- ◆ Seizures

## Signs and Symptoms

The cardinal sign of respiratory alkalosis is deep, rapid breathing, possibly exceeding 40 breaths/minute. This pattern of breathing is similar to Kussmaul's respirations that characterize diabetic acidosis. Such hyperventilation usually leads to CNS and neuromuscular disturbances, such as light-headedness or dizziness (because of below-normal CO<sub>2</sub> levels that decrease cerebral blood flow), agitation, circumoral and peripheral paresthesias, carpopedal spasms, twitching (possibly progressing to tetany), and muscle weakness. Severe respiratory alkalosis may cause cardiac arrhythmias (that may fail to respond to conventional treatment), seizures, or both.

## Diagnosis

 **CONFIRMING DIAGNOSIS** ABG analysis confirms respiratory alkalosis and rules out respiratory compensation for metabolic acidosis: Paco<sub>2</sub> less than 35 mm Hg; pH elevated in proportion to the fall in Paco<sub>2</sub> in the acute stage but falling toward normal in the chronic stage; and bicarbonate normal in the acute stage, but below normal in the chronic stage.

Chest X-ray or PFTs may aid in diagnosing possible lung disease.

## Treatment

Treatment is designed to eradicate the underlying condition—for example, removal of ingested toxins, treatment of fever or sepsis, providing oxygen for acute hypoxemia, and treatment of CNS disease. When hyperventilation is caused by severe anxiety, the patient may be instructed to breathe into a paper bag, which increases CO<sub>2</sub> levels and helps relieve anxiety.

Prevention of hyperventilation in patients receiving mechanical ventilation requires monitoring ABG levels and adjusting tidal volume and minute ventilation.

## Special Considerations

- ◆ Watch for and report any changes in neurologic, neuromuscular, or cardiovascular functions.
- ◆ Remember that twitching and cardiac arrhythmias may be associated with alkalemia and electrolyte imbalances. Monitor ABG and serum electrolyte levels closely, reporting any variations immediately.
- ◆ Explain all diagnostic tests and procedures to reduce anxiety.

## PNEUMOTHORAX

Pneumothorax is an accumulation of air or gas between the parietal and visceral pleurae. The amount of air or gas trapped in the intrapleural space determines the degree of lung collapse. In tension pneumothorax, the air in the pleural space is under higher pressure than air in adjacent lung and vascular structures. Without prompt treatment, tension or large pneumothorax results in fatal pulmonary and circulatory impairment. (See *Understanding tension pneumothorax*, page 112.)

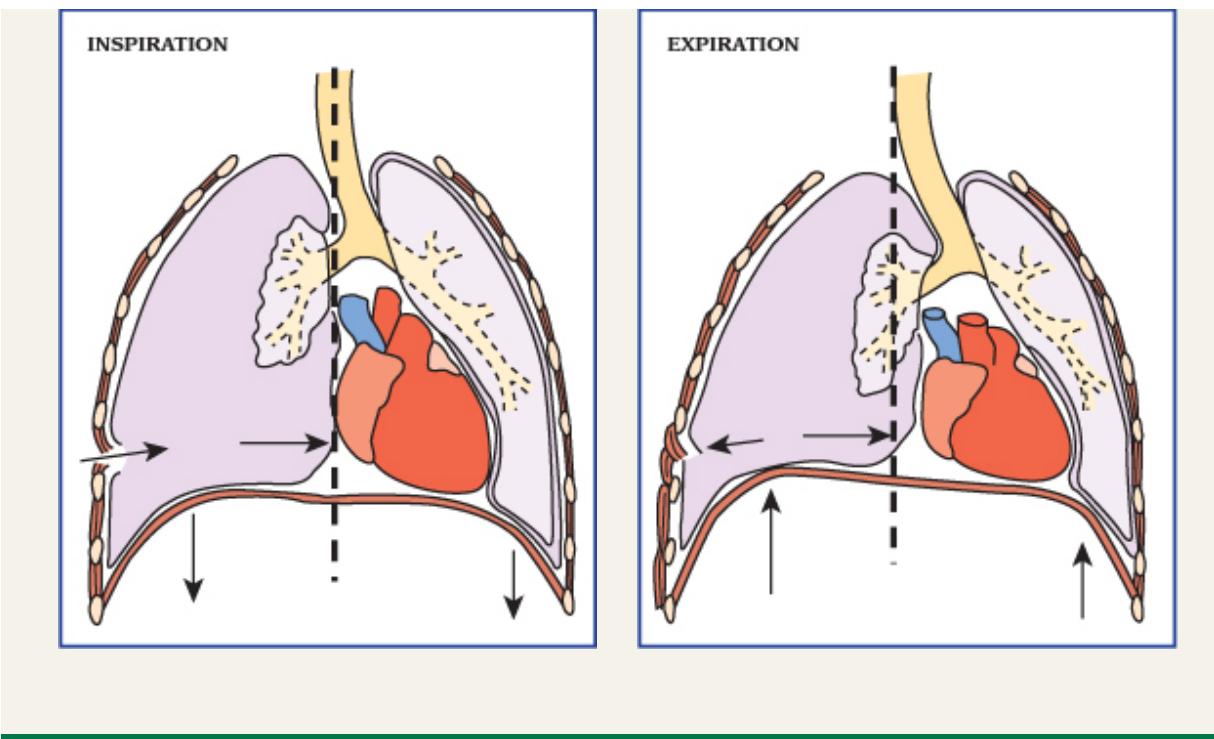


### PATHOPHYSIOLOGY UNDERSTANDING TENSION PNEUMOTHORAX

In tension pneumothorax, air accumulates intrapleurally and can't escape. Intrapleural pressure rises, collapsing the ipsilateral lung.

On inspiration, the mediastinum shifts toward the unaffected lung, impairing ventilation.

On expiration, the mediastinal shift distorts the vena cava and reduces venous return.



## Causes and Incidence

*Spontaneous pneumothorax* usually occurs in otherwise healthy adults 20 to 40 years old. It may be caused by air leakage from ruptured congenital blebs adjacent to the visceral pleural surface, near the apex of the lung. Secondary spontaneous pneumothorax is a complication of underlying lung disease, such as COPD, asthma, cystic fibrosis, TB, and whooping cough. Spontaneous pneumothorax may also occur in interstitial lung disease, such as eosinophilic granuloma or lymphangiomatosis.

*Traumatic pneumothorax* may result from insertion of a central venous line, thoracic surgery, or a penetrating chest injury, such as a gunshot or knife wound. It may follow a transbronchial biopsy, or it may also occur during thoracentesis or a closed pleural biopsy. When traumatic pneumothorax follows a penetrating chest injury, it frequently coexists with hemothorax (blood in the pleural space).

In *tension pneumothorax*, positive pleural pressure develops as a result of traumatic pneumothorax. When air enters the pleural space through a tear in lung tissue and is unable to leave by the same vent, each inspiration traps air in the pleural space, resulting in positive pleural pressure. This in turn causes collapse of the ipsilateral lung and marked impairment of venous return, which can severely compromise cardiac output and may cause a mediastinal

shift. Decreased filling of the great veins of the chest results in diminished cardiac output and lowered blood pressure.

## Pathophysiology

Pneumothorax can be classified as open or closed. In *open pneumothorax* (usually the result of trauma), air flows between the pleural space and the outside of the body. In *closed pneumothorax*, air reaches the pleural space directly from the lung.

## Complications

- ◆ Fatal pulmonary and circulatory impairment

## Signs and Symptoms

The cardinal features of pneumothorax are sudden, sharp, pleuritic pain (exacerbated by movement of the chest, breathing, and coughing); asymmetrical chest wall movement; and shortness of breath. Additional signs of tension pneumothorax are weak and rapid pulse, pallor, jugular vein distention, and anxiety. Tracheal deviations may be present with mediastinal shift. Tension pneumothorax produces the most severe respiratory symptoms; a spontaneous pneumothorax that releases only a small amount of air into the pleural space may cause no symptoms. In a nontension pneumothorax, the severity of symptoms is usually related to the size of the pneumothorax and the degree of preexisting respiratory disease.

## Diagnosis

Sudden, sharp chest pain and shortness of breath suggest pneumothorax.

 **CONFIRMING DIAGNOSIS** *Chest X-ray showing air in the pleural space and, possibly, mediastinal shift confirms this diagnosis.*

In the absence of a definitive chest X-ray, the physical examination may reveal:

- ◆ on inspection—overexpansion and rigidity of the affected chest side; in tension pneumothorax, jugular vein distention with hypotension and tachycardia
- ◆ on palpation—crackling beneath the skin, indicating subcutaneous emphysema (air in tissue) and decreased vocal fremitus

- ◆ on percussion—hyperresonance on the affected side
- ◆ on auscultation—decreased or absent breath sounds over the collapsed lung

If the pneumothorax is significant, ABG findings include pH less than 7.35,  $\text{Pao}_2$  less than 80 mm Hg, and  $\text{Paco}_2$  above 45 mm Hg.

## Treatment

Treatment is conservative for spontaneous pneumothorax in which no signs of increased pleural pressure (indicating tension pneumothorax) appear, lung collapse is less than 30%, and the patient shows no signs of dyspnea or other indications of physiologic compromise. Such treatment consists of bed rest, careful monitoring of blood pressure and pulse and respiratory rates, oxygen administration and, possibly, needle aspiration of air with a large-bore needle attached to a syringe. If more than 30% of the lung is collapsed, treatment to re-expand the lung includes placing a thoracostomy tube in the second or third intercostal space in the midclavicular line (or in the fifth or sixth intercostal space in the midaxillary line), connected to an underwater seal or low suction pressures.

Recurring spontaneous pneumothorax requires thoracotomy and pleurectomy; these procedures prevent recurrence by causing the lung to adhere to the parietal pleura. Traumatic and tension pneumothoraces require chest tube drainage; traumatic pneumothorax may also require surgery.

## Special Considerations



**ALERT** *Watch for pallor, gasping respirations, and sudden chest pain. Monitor patient's vital signs at least every hour for signs of shock, increasing respiratory distress, or mediastinal shift. Listen for breath sounds over both lungs. Falling blood pressure and rising pulse and respiratory rates may indicate tension pneumothorax, which can be fatal without prompt treatment.*

- ◆ Urge the patient to control coughing and gasping during thoracotomy. However, after the chest tube is in place, encourage them to cough and breathe deeply (at least once an hour) to facilitate lung expansion.
- ◆ If the patient is undergoing chest tube drainage, watch for continuing air leakage (bubbling), indicating the lung defect has failed to close; this may require surgery. Also watch for increasing subcutaneous emphysema by

checking around the neck or at the tube insertion site for crackling beneath the skin. If the patient is on a ventilator, watch for difficulty in breathing in time with the ventilator as well as pressure changes on ventilator gauges.

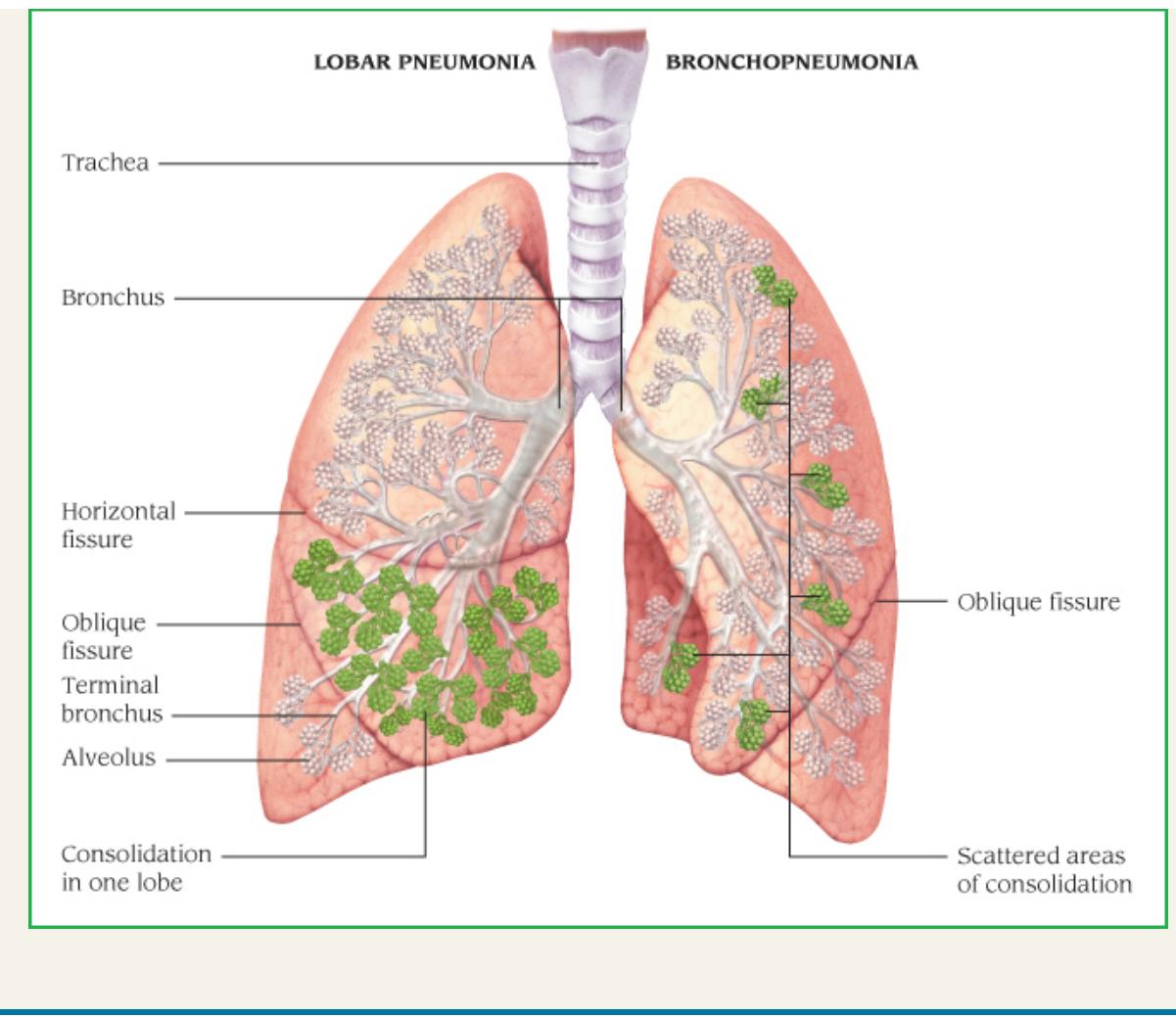
- ◆ Change dressings around the chest tube insertion site according to your facility's policy. Don't reposition or dislodge the tube. If it dislodges, immediately place a petroleum gauze dressing over the opening to prevent rapid lung collapse.
- ◆ Secure the chest tube drainage apparatus appropriately. Tape connections securely.
- ◆ Monitor the patient's vital signs frequently after thoracotomy. Also, for the first 24 hours, assess respiratory status by checking breath sounds hourly. Observe the chest tube site for leakage, noting the amount and color of drainage. Help the patient walk, as ordered (usually on the first postoperative day), to facilitate deep inspiration and lung expansion.
- ◆ To reassure the patient, explain what pneumothorax is, what causes it, and all diagnostic tests and procedures. Make them as comfortable as possible. (The patient with pneumothorax is usually most comfortable sitting upright.)

## PNEUMONIA

Pneumonia is an acute infection of the lung parenchyma that commonly impairs gas exchange. The prognosis is generally good for people who have normal lungs and adequate host defenses before the onset of pneumonia; however, pneumonia is the sixth leading cause of death in the United States. (See *Looking at lobar pneumonia and bronchopneumonia*.)

### Looking at Lobar Pneumonia and Bronchopneumonia

Pneumonia can involve the distal airways, alveoli, part of a lobe, or an entire lobe.



## Causes and Incidence

Pneumonia can be classified in several ways:

- ◆ Microbiologic etiology—Pneumonia can be viral, bacterial, fungal, protozoan, mycobacterial, mycoplasmal, or rickettsial in origin. (See *Diagnosing and treating the types of pneumonia*, pages 114 to 116.)

## Diagnosing and Treating the Types of Pneumonia

| Type | Signs and symptoms | Diagnosis | Treatment |
|------|--------------------|-----------|-----------|
|------|--------------------|-----------|-----------|

| Type  | Signs and symptoms   | Diagnosis   | Treatment  |
|---|--|---|--|
| <b>Aspiration</b><br>Results from vomiting and aspiration of gastric or oropharyngeal contents into trachea and lungs | <ul style="list-style-type: none"> <li>◆ Noncardiogenic pulmonary edema that may follow damage to respiratory epithelium from contact with stomach acid</li> <li>◆ Crackles, dyspnea, cyanosis, hypotension, and tachycardia</li> <li>◆ May be subacute pneumonia with cavity formation; lung abscess may occur if foreign body is present</li> </ul>  | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray</i>: locates areas of infiltrates, which suggest diagnosis</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Antimicrobial therapy</i>: penicillin G or clindamycin</li> <li>◆ <i>Supportive</i>: oxygen therapy, suctioning, coughing, deep breathing, and adequate hydration</li> </ul>       |
| <b>Bacterial</b><br><i>Klebsiella</i>   | <ul style="list-style-type: none"> <li>◆ Fever and recurrent chills; cough producing rusty, bloody, viscous sputum (currant jelly); cyanosis of lips and nail beds due to hypoxemia; and shallow, grunting respirations</li> <li>◆ Common in patients with chronic alcoholism, pulmonary disease, diabetes, or those at risk for aspiration</li> </ul> | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray</i>: typically, but not always, consolidation in the upper lobe that causes bulging of fissures</li> <li>◆ <i>White blood cell (WBC) count</i>: elevated</li> <li>◆ <i>Sputum culture and Gram stain</i>: may show gram-negative <i>Klebsiella</i></li> </ul> | <ul style="list-style-type: none"> <li>◆ <i>Antimicrobial therapy</i>: an aminoglycoside and a cephalosporin</li> </ul>  |
| <i>Staphylococcus</i>   | <ul style="list-style-type: none"> <li>◆ Temperature of 102° to 104° F (38.9° to 40° C), recurrent shaking chills, bloody sputum, dyspnea, tachypnea, and hypoxemia</li> <li>◆ Should be suspected with viral illness, such as influenza or measles, and in patients with cystic fibrosis</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray</i>: multiple abscesses and infiltrates; high incidence of empyema</li> <li>◆ <i>WBC count</i>: elevated</li> <li>◆ <i>Sputum culture and Gram stain</i>: may show gram-positive staphylococci</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Antimicrobial therapy</i>: nafcillin or oxacillin for 14 days if staphylococci are penicillinase producing</li> <li>◆ <i>Supportive</i>: chest tube drainage of empyema</li> </ul> |

| Type  | Signs and symptoms   | Diagnosis   | Treatment  |
|---|--|---|--|
| Streptococcus<br>( <i>Streptococcus pneumoniae</i> )                            | Sudden onset of severe, shaking chills and a sustained temperature of 102° to 104° F (38.9° to 40° C); commonly preceded by upper respiratory tract infection  | <ul style="list-style-type: none"> <li>◆ Chest X-ray: areas of consolidation, commonly lobar</li> <li>◆ WBC count: elevated</li> <li>◆ Sputum culture: may show gram-positive <i>S. pneumoniae</i>; this organism not always recovered</li> </ul> | <ul style="list-style-type: none"> <li>◆ Antimicrobial therapy: penicillin G (or erythromycin, if patient is allergic to penicillin) for 7 to 10 days beginning after obtaining culture specimen but without waiting for results. (Resistance to penicillin is becoming much more common and, in the patient with risk factors for resistance [extreme age, day care attendance, or immunosuppression], treatment with vancomycin, imipenem, or levofloxacin should be considered.)</li> </ul> |
| <b>Protozoan</b><br><i>Pneumocystis carinii</i> (jiroveci)                      | <ul style="list-style-type: none"> <li>◆ Occurs in immunocompromised persons</li> <li>◆ Dyspnea and nonproductive cough</li> <li>◆ Anorexia, weight loss, and fatigue</li> <li>◆ Low-grade fever</li> </ul>              | <ul style="list-style-type: none"> <li>◆ Fiber-optic bronchoscopy: obtains specimens for histologic studies</li> <li>◆ Chest X-ray: nonspecific infiltrates, nodular lesions, or spontaneous pneumothorax</li> </ul>                              | <ul style="list-style-type: none"> <li>◆ Antimicrobial therapy: trimethoprim and sulfamethoxazole or pentamidine by I.V. administration or inhalation. (Prophylactic pentamidine may be used for high-risk patients.)</li> <li>◆ Supportive: oxygen, improved nutrition, and mechanical ventilation</li> </ul>   |
| <b>Viral</b><br>Adenovirus<br>(insidious onset; generally affects young adults) | <ul style="list-style-type: none"> <li>◆ Sore throat, fever, cough, chills, malaise, small amounts of mucoid sputum, retrosternal chest pain, anorexia, rhinitis, adenopathy, scattered crackles, and rhonchi</li> </ul> | <ul style="list-style-type: none"> <li>◆ Chest X-ray: patchy distribution of pneumonia, more severe than indicated by physical examination</li> <li>◆ WBC count: normal to slightly elevated</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Treat symptoms only</li> <li>◆ Mortality low; usually clears with no residual effects</li> </ul>  |

| Type  | Signs and symptoms   | Diagnosis  | Treatment   |
|---|--|--|---|
| Chicken pox<br>(varicella)<br>(uncommon in children, but present in 30% of adults with varicella) | <ul style="list-style-type: none"> <li>◆ Cough, dyspnea, cyanosis, tachypnea, pleuritic chest pain, hemoptysis, and rhonchi 1 to 6 days after onset of rash</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray:</i> shows more extensive pneumonia than indicated by physical examination and bilateral, patchy, diffuse, nodular infiltrates</li> <li>◆ <i>Sputum analysis:</i> predominant mononuclear cells and characteristic intranuclear inclusion bodies, with characteristic skin rash, confirm diagnosis</li> </ul>  | <ul style="list-style-type: none"> <li>◆ <i>Supportive:</i> adequate hydration, and oxygen therapy in critically ill patients</li> <li>◆ Therapy with I.V. acyclovir</li> </ul>   |
| Viral<br>Cytomegalovirus  | <ul style="list-style-type: none"> <li>◆ Difficult to distinguish from other nonbacterial pneumonias</li> <li>◆ Fever, cough, shaking chills, dyspnea, cyanosis, weakness, and diffuse crackles</li> <li>◆ Occurs in neonates as devastating multisystemic infection; in normal adults, resembles mononucleosis; in immunocompromised hosts, varies from clinically inapparent to devastating infection</li> </ul> | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray:</i> in early stages, variable patchy infiltrates; later, bilateral, nodular, and more predominant in lower lobes</li> <li>◆ <i>Percutaneous aspiration of lung tissue, transbronchial biopsy, or open lung biopsy:</i> microscopic examination shows typical intranuclear and cytoplasmic inclusions; the virus can be cultured from lung tissue</li> </ul> | <ul style="list-style-type: none"> <li>◆ Generally, benign and self-limiting in mononucleosis-like form</li> <li>◆ <i>Supportive:</i> adequate hydration and nutrition, oxygen therapy, and bed rest</li> <li>◆ In immunosuppressed patients, disease is more severe and may be fatal; ganciclovir or foscarnet treatment is warranted</li> </ul> |

| Type  | Signs and symptoms  | Diagnosis   | Treatment   |
|---|---|---|---|
| Influenza<br>(prognosis poor even with treatment; 30% mortality)        | <ul style="list-style-type: none"> <li>◆ Cough (initially nonproductive; later, purulent sputum), marked cyanosis, dyspnea, high fever, chills, substernal pain and discomfort, moist crackles, frontal headache, and myalgia</li> <li>◆ Death results from cardiopulmonary collapse</li> </ul> | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray:</i> diffuse bilateral bronchopneumonia radiating from hilus</li> <li>◆ <i>WBC count:</i> normal to slightly elevated</li> <li>◆ <i>Sputum smears:</i> no specific organisms</li> </ul> | <ul style="list-style-type: none"> <li>◆ <i>Supportive:</i> for respiratory failure, endotracheal intubation and ventilator assistance; for fever, hypothermia blanket or antipyretics; and for influenza A, amantadine or rimantadine</li> </ul> |
| Measles<br>(rubeola)  | <ul style="list-style-type: none"> <li>◆ Fever, dyspnea, cough, small amounts of sputum, coryza, rash, and cervical adenopathy</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray:</i> reticular infiltrates, sometimes with hilar lymph node enlargement</li> <li>◆ <i>Lung tissue specimen:</i> characteristic giant cells</li> </ul>                                   | <ul style="list-style-type: none"> <li>◆ <i>Supportive:</i> bed rest, adequate hydration, and antimicrobials; assisted ventilation if necessary</li> </ul>  |
| Respiratory syncytial virus<br>(most prevalent in infants and children) | <ul style="list-style-type: none"> <li>◆ Listlessness, irritability, tachypnea with retraction of intercostal muscles, wheezing, slight sputum production, fine moist crackles, fever, severe malaise, and cough</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Chest X-ray:</i> patchy bilateral consolidation</li> <li>◆ <i>WBC count:</i> normal to slightly elevated</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Supportive:</i> humidified air, oxygen, antimicrobials (commonly given until viral etiology confirmed), and aerosolized ribavirin</li> <li>◆ Usually complete recovery</li> </ul>                     |

- ◆ Location—Bronchopneumonia involves distal airways and alveoli; lobular pneumonia, part of a lobe; and lobar pneumonia, an entire lobe.
- ◆ Type—Primary pneumonia results from inhalation or aspiration of a pathogen; it includes pneumococcal and viral pneumonia. Secondary pneumonia may follow initial lung damage from a noxious chemical or other insult (superinfection) or may result from hematogenous spread of bacteria from a distant focus.

Predisposing factors for bacterial and viral pneumonia include chronic illness and debilitation, cancer (particularly lung cancer), abdominal and thoracic surgery, atelectasis, common colds or other viral respiratory

infections, such as acquired immunodeficiency syndrome, chronic respiratory disease (COPD, asthma, bronchiectasis, and cystic fibrosis), influenza, smoking, malnutrition, alcoholism, sickle cell disease, tracheostomy, exposure to noxious gases, aspiration, and immunosuppressive therapy.

Predisposing factors for aspiration pneumonia include old age, debilitation, artificial airway use, NG tube feedings, impaired gag reflex, poor oral hygiene, and decreased level of consciousness.

In elderly patients and patients who are debilitated, bacterial pneumonia may follow influenza or a common cold. Respiratory viruses are the most common cause of pneumonia in children 2 to 3 years old. In school-age children, mycoplasma pneumonia is more common.

## **Pathophysiology**

A pathogen or extrinsic agent enters the respiratory tract and invades or insults the alveoli. When the host defenses are overwhelmed by the inoculum size or virulence of the pathogen, it results in an infection of the lung parenchyma.

## **Complications**

- ◆ Septic shock
- ◆ Hypoxemia
- ◆ Respiratory failure
- ◆ Empyema
- ◆ Lung abscess
- ◆ Bacteremia
- ◆ Endocarditis
- ◆ Pericarditis
- ◆ Meningitis

## **Signs and Symptoms**

The main symptoms of pneumonia are coughing, sputum production, pleuritic chest pain, shaking chills, shortness of breath, rapid shallow breathing, and fever. Physical signs vary widely, ranging from diffuse, fine crackles to signs of localized or extensive consolidation and pleural effusion. There may also be associated symptoms of headache, sweating, loss of appetite, excess fatigue, and confusion (in older people).

Complications include hypoxemia, respiratory failure, pleural effusion, empyema, lung abscess, and bacteremia, with spread of infection to other parts of the body, resulting in meningitis, endocarditis, and pericarditis.

## Diagnosis

Clinical features, chest X-ray showing infiltrates, and sputum smear demonstrating acute inflammatory cells support the diagnosis. Gram stain and sputum culture may identify the organism. Positive blood cultures in the patient with pulmonary infiltrates strongly suggest pneumonia produced by the organisms isolated from the blood cultures. Pleural effusions, if present, should be tapped and fluid analyzed for evidence of infection in the pleural space. Occasionally, a transtracheal aspirate of tracheobronchial secretions or bronchoscopy with brushings or washings may be done to obtain material for smear and culture. The patient's response to antimicrobial therapy also provides important evidence of the presence of pneumonia.

## Treatment

Antimicrobial therapy varies with the causative agent. Therapy should be reevaluated early in the course of treatment. Supportive measures include humidified oxygen therapy for hypoxemia, mechanical ventilation for respiratory failure, a high-calorie diet and adequate fluid intake, bed rest, and an analgesic to relieve pleuritic chest pain. Patients with severe pneumonia on mechanical ventilation may require PEEP to facilitate adequate oxygenation.

## Special Considerations

Correct supportive care can increase patient comfort, avoid complications, and speed recovery.

The following protocol should be observed throughout the illness:

- ◆ Maintain a patent airway and adequate oxygenation. Monitor pulse oximetry. Measure ABG levels, especially in hypoxic patients. Administer supplemental oxygen if the  $\text{PaO}_2$  is less than 55 to 60 mm Hg. Patients with underlying chronic lung disease should be given oxygen cautiously.
- ◆ Teach the patient how to cough and perform deep-breathing exercises to clear secretions; encourage them to do so often. In severe pneumonia that requires ET intubation or tracheostomy (with or without mechanical

ventilation), provide thorough respiratory care. Suction often, using sterile technique, to remove secretions.

- ◆ Obtain sputum specimens as needed, by suction if the patient can't produce specimens independently. Collect specimens in a sterile container and deliver them promptly to the microbiology laboratory.
- ◆ Administer antibiotics as ordered, sedation, and pain medication as needed; record the patient's response to medications. Fever and dehydration may require I.V. fluids and electrolyte replacement.
- ◆ Maintain adequate nutrition to offset hypermetabolic state secondary to infection. Ask the dietary department to provide a high-calorie, high-protein diet consisting of soft, easy-to-eat foods. Encourage the patient to eat. As necessary, supplement oral feedings with NG tube feedings or parenteral nutrition. Monitor fluid intake and output. Consider limiting the use of milk products because they may increase sputum production.
- ◆ Provide a quiet, calm environment for the patient, with frequent rest periods.
- ◆ Give emotional support by explaining all procedures (especially intubation and suctioning) to the patient and family. Encourage family visits. Provide diversionary activities appropriate to the patient's age.
- ◆ To control the spread of infection, dispose of secretions properly. Tell the patient to sneeze and cough into a disposable tissue; tape a lined bag to the side of the bed for used tissues.

Pneumonia can be prevented as follows:

- ◆ Advise the patient to avoid using antibiotics indiscriminately during minor viral infections because this may result in upper airway colonization with antibiotic-resistant bacteria. If the patient then develops pneumonia, the organisms producing the pneumonia may require treatment with more toxic antibiotics.
- ◆ Encourage pneumococcal vaccine (pneumovax) and annual influenza vaccination for high-risk patients, such as those with COPD, chronic heart disease, or sickle cell disease.
- ◆ Urge all bedridden and postoperative patients to perform deep-breathing and coughing exercises frequently. Reposition such patients often to promote full aeration and drainage of secretions. Encourage early ambulation in postoperative patients.
- ◆ To prevent aspiration during NG tube feedings, elevate the patient's head, check the tube's position, and administer the formula slowly. Don't give

large volumes at one time; this could cause vomiting. Keep the patient's head elevated for at least 30 minutes after the feeding. Check for residual formula at 4- to 6-hour intervals.



## PREVENTION

- *Perform hand hygiene frequently to prevent infections.*
- *Advise patients to avoid taking antibiotics indiscriminately during viral infections.*

## IDIOPATHIC BRONCHIOLITIS OBLITERANS WITH ORGANIZING PNEUMONIA

Idiopathic bronchiolitis obliterans with organizing pneumonia (BOOP), also known as *cryptogenic organizing pneumonia*, is one of several types of bronchiolitis obliterans. *Organizing pneumonia* refers to unresolved pneumonia, in which inflammatory alveolar exudate persists and eventually undergoes fibrosis. *Bronchiolitis obliterans* is a generic term used to describe an inflammatory disease of the small airways.

### Causes and Incidence

BOOP has no known cause. However, other forms of bronchiolitis obliterans and organizing pneumonia may be associated with specific diseases or situations, such as bone marrow, heart, or heart-lung transplantation; collagen vascular diseases, such as rheumatoid arthritis and systemic lupus erythematosus (LE); inflammatory diseases, such as Crohn disease, ulcerative colitis, and polyarteritis nodosa; bacterial, viral, or mycoplasmal respiratory infections; inhalation of toxic gases; and drug therapy with amiodarone, bleomycin, penicillamine, or lomustine.

Much debate still exists about the various pathologies and classifications of bronchiolitis obliterans. Most patients with BOOP are between 50 and 60 years old. Incidence is equally divided between men and women. A smoking history doesn't seem to increase the risk of developing BOOP.

### Pathophysiology

BOOP is the result of an epithelial injury in the lung that progresses to the formation of fibrinoid inflammatory cell clusters and subsequently intra-

alveolar fibrosis. Upon biopsy the key characteristic is fibroblastic plugs present in the alveoli, alveolar ducts, and bronchioles.

## Complications

- ◆ Respiratory failure
- ◆ Interstitial lung disease
- ◆ Death

## Signs and Symptoms

The presenting symptoms of BOOP are usually subacute, with a flulike syndrome of fever, persistent and nonproductive cough, dyspnea (especially with exertion), malaise, anorexia, and weight loss lasting for several weeks to several months. Physical assessment findings may reveal dry crackles as the only abnormality. Less common symptoms include a productive cough, hemoptysis, chest pain, generalized aching, and night sweats.

## Diagnosis

Diagnosis begins with a thorough patient history meant to exclude any known cause of bronchiolitis obliterans or diseases with a pathophysiology that includes an organizing pneumonia pattern.

- ◆ Chest X-ray usually shows patchy, diffuse airspace opacities with a ground-glass appearance that may migrate from one location to another. High-resolution CT scans show areas of consolidation. Except for the migrating opacities, these findings are nonspecific and present in many other respiratory disorders.
- ◆ PFTs may be normal or show reduced capacities. The diffusing capacity for carbon monoxide is generally low.
- ◆ ABG analysis usually shows mild to moderate hypoxemia at rest, which worsens with exercise.
- ◆ Blood tests reveal an increased erythrocyte sedimentation rate, an increased C-reactive protein level, and an increased WBC count with a somewhat increased proportion of neutrophils and a minor rise in eosinophils. Immunoglobulin (Ig) G and IgM levels are normal or slightly increased, and the IgE level is normal.
- ◆ Bronchoscopy reveals normal or slightly inflamed airways. Bronchoalveolar lavage fluid obtained during bronchoscopy shows a moderate elevation in lymphocytes and, sometimes, elevated neutrophil

and eosinophil levels. Foamy-looking alveolar macrophages may also be found.

**D** **CONFIRMING DIAGNOSIS** *Lung biopsy, thoracoscopy, or bronchoscopy is required to confirm the diagnosis of BOOP. Pathologic changes in lung tissue include plugs of connective tissue in the lumen of the bronchioles, alveolar ducts, and alveolar spaces.*

These changes may occur in other types of bronchiolitis and in other diseases that cause organizing pneumonia. They also differentiate BOOP from constrictive bronchiolitis (characterized by inflammation and fibrosis that surrounds and may narrow or completely obliterate the bronchiolar airways). Although the pathologic findings in proliferative and constrictive bronchiolitis are different, the causes and presentations may overlap. Any known cause of bronchiolitis obliterans or organizing pneumonia must be ruled out before the diagnosis of BOOP is made.

## Treatment

Corticosteroids are the current treatment for BOOP, although the ideal dosage and duration of treatment remain topics of discussion. Relapse is common when steroids are tapered off or stopped. This usually can be reversed when steroids are increased or resumed. Occasionally, a patient may need to continue corticosteroids indefinitely.

Immunosuppressive-cytotoxic drugs, such as cyclophosphamide, have been used in the few cases of intolerance or unresponsiveness.

Oxygen is used to correct hypoxemia. The patient may need either no oxygen or a small amount of oxygen at rest and a greater amount when they exercise.

Other treatments vary, depending on the patient's symptoms, and may include inhaled bronchodilators, cough suppressants, and bronchial hygiene therapies.

BOOP is very responsive to treatment and usually can be completely reversed with corticosteroid therapy. However, a few deaths have been reported, particularly in patients who had more widespread pathologic changes in the lung or patients who developed opportunistic infections or other complications related to steroid therapy.

## Special Considerations

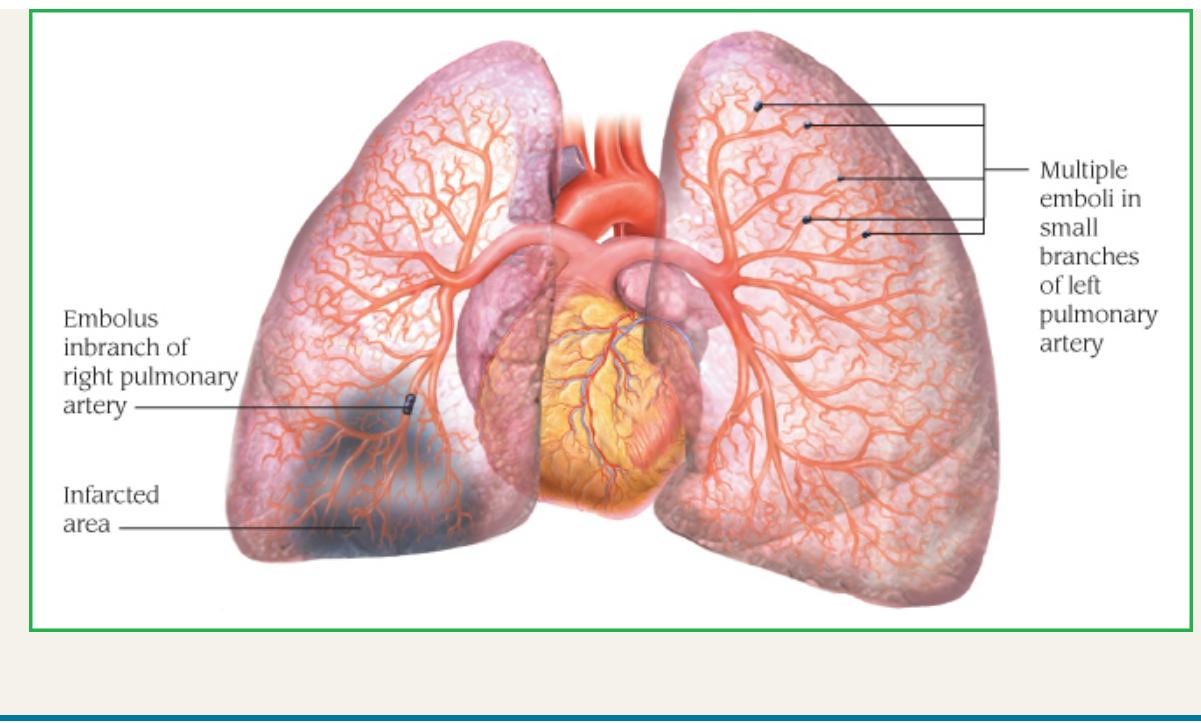
- ◆ Explain all diagnostic tests. The patient may experience anxiety and frustration because of the length of time and number of tests needed to establish the diagnosis.
- ◆ Explain the diagnosis to the patient and family. This uncommon diagnosis may cause confusion and anxiety.
- ◆ Monitor the patient for adverse effects of corticosteroid therapy: weight gain, “moon face,” glucose intolerance, fluid and electrolyte imbalance, mood swings, cataracts, peptic ulcer disease, opportunistic infections, and osteoporosis leading to bone fractures. In many cases, these effects leave the patient unable to tolerate the treatment. Teach the patient and their family about these adverse effects, emphasizing which reactions should be reported to the physician.
- ◆ Teach measures that may help prevent complications related to treatment, such as infection control and improved nutrition.
- ◆ Teach breathing, relaxation, and energy conservation techniques to help the patient manage symptoms.
- ◆ Monitor oxygenation, both at rest and with exertion. The physician will probably prescribe an oxygen flow rate for use when the patient is at rest and a higher one for exertion. Teach the patient how to increase the oxygen flow rate to the appropriate level for exercise.
- ◆ If the patient needs oxygen at home, ensure continuity of care by making appropriate referrals to discharge planners, respiratory care practitioners, and home equipment vendors.

## PULMONARY EMBOLISM

The most common pulmonary complication in hospitalized patients, pulmonary embolism is an obstruction of the pulmonary arterial bed by a dislodged thrombus, heart valve vegetation, or foreign substance. Although pulmonary infarction that results from embolism may be so mild as to be asymptomatic, massive embolism (more than 50% obstruction of pulmonary arterial circulation) and the accompanying infarction can be rapidly fatal. (See *Looking at pulmonary emboli*, page 120.)

### Looking at Pulmonary Emboli

This illustration shows multiple emboli in pulmonary artery branches and a larger embolus that has resulted in an infarcted area in the lung.



## Causes and Incidence

Pulmonary embolism generally results from dislodged thrombi originating in the leg veins. More than half of such thrombi arise in the deep veins of the legs. Other less common sources of thrombi are the pelvic veins, renal veins, hepatic vein, right side of the heart, and upper extremities. Such thrombus formation results directly from vascular wall damage, venostasis, or hypercoagulability of the blood. Trauma, clot dissolution, sudden muscle spasm, intravascular pressure changes, or a change in peripheral blood flow can cause the thrombus to loosen or fragment. Then the thrombus—now called an *embolus*—floats to the heart's right side and enters the lung through the pulmonary artery. There, the embolus may dissolve, continue to fragment, or grow.

By occluding the pulmonary artery, the embolus prevents alveoli from producing enough surfactant to maintain alveolar integrity. As a result, alveoli collapse and atelectasis develops. If the embolus enlarges, it may clog most or all of the pulmonary vessels and cause death.

Rarely, the emboli contain air, fat, bacteria, amniotic fluid, talc (from drugs intended for oral administration, which are injected intravenously by addicts), or tumor cells.

Predisposing factors for pulmonary embolism include long-term immobility, chronic pulmonary disease, heart failure or atrial fibrillation,

thrombophlebitis, polycythemia vera, thrombocytosis, autoimmune hemolytic anemia, sickle cell disease, varicose veins, recent surgery, advanced age, pregnancy, lower extremity fractures or surgery, burns, obesity, vascular injury, cancer, I.V. drug abuse, or hormonal contraceptives.

## **Pathophysiology**

Once a thrombus dislodges, it travels through to the venous system, through the right side of the heart, and lodges in the pulmonary arteries. The thrombus can completely occlude the vessels or only partially block the flow of blood. Many physiologic factors participate in the potential outcomes including the ability of the body's thrombolytic system to break down clots, function of the right ventricle, over all condition of lungs, and number and size of emboli.

## **Complications**

- ◆ Pulmonary infarction
- ◆ Death

## **Signs and Symptoms**

Total occlusion of the main pulmonary artery is rapidly fatal; smaller or fragmented emboli produce symptoms that vary with the size, number, and location of the emboli. Usually, the first symptom of pulmonary embolism is dyspnea, which may be accompanied by anginal or pleuritic chest pain. Other clinical features include tachycardia, productive cough (sputum may be blood-tinged), low-grade fever, and pleural effusion. Less common signs include massive hemoptysis, chest splinting, leg edema and, with a large embolus, cyanosis, syncope, and distended jugular veins.

In addition, pulmonary embolism may cause pleural friction rub and signs of circulatory collapse (weak, rapid pulse, and hypotension) and hypoxia (restlessness and anxiety).

## **Diagnosis**

The patient history should reveal predisposing conditions for pulmonary embolism. A triad of deep vein thrombosis (DVT) formation is stasis, endothelial injury, and hypercoagulability. Risk factors include long car or plane trips, cancer, pregnancy, hypercoagulability, prior DVT, and pulmonary emboli.

- ◆ Chest X-ray helps to rule out other pulmonary diseases; areas of atelectasis, an elevated diaphragm and pleural effusion, a prominent pulmonary artery and, occasionally, the characteristic wedge-shaped infiltrate suggestive of pulmonary infarction, or focal oligemia of blood vessels, are apparent.
- ◆ Lung scan shows perfusion defects in areas beyond occluded vessels; however, it doesn't rule out microemboli.
- ◆ Pulmonary angiography is the most definitive test but requires a skilled angiographer and radiologic equipment; it also poses some risk to the patient. Its use depends on the uncertainty of the diagnosis and the need to avoid unnecessary anticoagulant therapy in a high-risk patient.
- ◆ Electrocardiography may show right axis deviation; right bundle-branch block; tall, peaked P waves; ST-segment depression and T-wave inversions (indicative of right-sided heart strain); and supraventricular tachyarrhythmias in extensive pulmonary embolism. A pattern sometimes observed is S<sub>1</sub>, Q<sub>3</sub>, and T<sub>3</sub> (S wave in lead I, Q wave in lead III, and inverted T wave in lead III).
- ◆ Auscultation occasionally reveals a right ventricular S<sub>3</sub> gallop and increased intensity of the pulmonic component of S<sub>2</sub>. Also, crackles and a pleural rub may be heard at the embolism site.
- ◆ ABG analysis showing a decreased Pao<sub>2</sub> and Paco<sub>2</sub> are characteristic but don't always occur.

If pleural effusion is present, thoracentesis may rule out empyema, which indicates pneumonia.

## Treatment

Treatment is designed to maintain adequate cardiovascular and pulmonary function during resolution of the obstruction and to prevent recurrence of embolic episodes. Because most emboli resolve within 10 to 14 days, treatment consists of oxygen therapy as needed and anticoagulation with heparin to inhibit new thrombus formation, followed by oral warfarin. Heparin therapy is monitored by daily coagulation studies (PTT).

Patients with massive pulmonary embolism and shock may need fibrinolytic therapy with thrombolytic therapy (streptokinase, urokinase, or tissue plasminogen activator) to enhance fibrinolysis of the pulmonary emboli and remaining thrombi. Emboli that cause hypotension may require the use of vasopressors. Treatment of septic emboli requires antibiotics—not

anticoagulants—and evaluation for the infection's source, particularly endocarditis.

Surgery is performed on patients who can't take anticoagulants, who have recurrent emboli during anticoagulant therapy, or who have been treated with thrombolytic agents or pulmonary thromboendarterectomy. This procedure (which shouldn't be performed without angiographic evidence of pulmonary embolism) consists of vena cava ligation, plication, or insertion of an inferior vena cava device to filter blood returning to the heart and lungs.

## Special Considerations

- ◆ Give oxygen by nasal cannula or mask. Check ABG levels if the patient develops fresh emboli or worsening dyspnea. Be prepared to provide ET intubation with assisted ventilation if breathing is severely compromised.
- ◆ Administer heparin, as ordered, through I.V. push or continuous drip. Monitor coagulation studies daily. Effective heparin therapy raises the PTT to more than 1½ times normal. Watch closely for nosebleeds, petechiae, and other signs of abnormal bleeding; check stools for occult blood. Patients should be protected from trauma and injury; avoid I.M. injections and maintain pressure over venipuncture sites for 5 minutes, or until bleeding stops, to reduce hematoma.
- ◆ After the patient is stable, encourage them to move about often, and assist with isometric and range-of-motion exercises. Check pedal pulses, temperature, and color of feet to detect venostasis. *Never* massage the patient's legs. Offer diversional activities to promote rest and relieve restlessness.
- ◆ Help the patient walk as soon as possible after surgery to prevent venostasis.
- ◆ Maintain adequate nutrition and fluid balance to promote healing.
- ◆ Report frequent pleuritic chest pain, so that analgesics can be prescribed. Also, incentive spirometry can assist in deep breathing. Provide tissues and a bag for easy disposal of expectorations.
- ◆ Warn the patient not to cross their legs; this promotes thrombus formation.
- ◆ To relieve anxiety, explain procedures and treatments. Encourage the patient's family to participate in their care.
- ◆ Most patients need treatment with an oral anticoagulant (warfarin) for 3 to 6 months after a pulmonary embolism. Advise these patients to watch for signs of bleeding (bloody stools, blood in urine, and large ecchymoses), to take the prescribed medication exactly as ordered, not to change dosages

without consulting their physician, and to avoid taking additional medication (including aspirin and vitamins). Stress the importance of follow-up laboratory tests (international normalized ratio) to monitor anticoagulant therapy.



## PREVENTION

- *Encourage early ambulation in patients predisposed to this condition. With close medical supervision, low-dose heparin may be useful prophylactically.*
- *In high-risk patients, low-molecular-weight heparin may be given.*

## SARCOIDOSIS

Sarcoidosis is a multisystem, granulomatous disorder that characteristically produces lymphadenopathy, pulmonary infiltration, and skeletal, liver, eye, or skin lesions. Acute sarcoidosis usually resolves within 2 years. Chronic, progressive sarcoidosis, which is uncommon, is associated with pulmonary fibrosis and progressive pulmonary disability.

### Causes and Incidence

The cause of sarcoidosis is unknown, but these factors may play a role:

- ◆ hypersensitivity response (possibly from T-cell imbalance) to such agents as atypical mycobacteria, fungi, and pine pollen
- ◆ genetic predisposition (suggested by a slightly higher incidence of sarcoidosis within the same family)
- ◆ extreme immune response to infection

Sarcoidosis occurs most commonly in adults 30 to 50 years old. In the United States, sarcoidosis occurs predominantly among blacks, affecting twice as many women as men.

### Pathophysiology

In the development of sarcoidosis, T cells play an important role by initiating a significant immune reaction. In sites where there is disease activity, a concentration of CD4 cells exist along with a release of interleukin-2. This results in a disequilibrium of the CD4/CD8 ratio. The exaggerated immune

response creates the characteristic noncaseating granulomas, found primarily on the lungs and intrathoracic lymph nodes.

## Complications

- ◆ Pulmonary fibrosis
- ◆ Pulmonary hypertension
- ◆ Cor pulmonale

## Signs and Symptoms

Initial symptoms of sarcoidosis include arthralgia (in the wrists, ankles, and elbows), fatigue, malaise, and weight loss. Other clinical features vary according to the extent and location of the fibrosis:

- ◆ Respiratory—breathlessness, cough (usually nonproductive), substernal pain; complications in advanced pulmonary disease include pulmonary hypertension and cor pulmonale
- ◆ Cutaneous—erythema nodosum, subcutaneous skin nodules with maculopapular eruptions, and extensive nasal mucosal lesions
- ◆ Ophthalmic—anterior uveitis (common), glaucoma, and blindness (rare)
- ◆ Lymphatic—bilateral hilar and right paratracheal lymphadenopathy and splenomegaly
- ◆ Musculoskeletal—muscle weakness, polyarthralgia, pain, and punched-out lesions on phalanges
- ◆ Hepatic—granulomatous hepatitis, usually asymptomatic
- ◆ Genitourinary—hypercalciuria
- ◆ Cardiovascular—arrhythmias (premature beats, bundle-branch or complete heart block) and, rarely, cardiomyopathy
- ◆ CNS—cranial or peripheral nerve palsies, basilar meningitis, seizures, and pituitary and hypothalamic lesions producing diabetes insipidus

## Diagnosis

Typical clinical features with appropriate laboratory data and X-ray findings suggest sarcoidosis. A positive skin lesion biopsy supports the diagnosis.

Other relevant findings include:

- ◆ Chest X-ray—bilateral hilar and right paratracheal adenopathy with or without diffuse interstitial infiltrates; occasionally large nodular lesions present in lung parenchyma

- ◆ Lymph node or lung biopsy—noncaseating granulomas with negative cultures for mycobacteria and fungi
- ◆ Other laboratory data—rarely, increased serum calcium, mild anemia, leukocytosis, and hyperglobulinemia
- ◆ PFTs—decreased TLC and compliance, and decreased diffusing capacity
- ◆ ABG analysis—decreased arterial oxygen tension

Negative tuberculin skin test, fungal serologies, and sputum cultures for mycobacteria and fungi as well as negative biopsy cultures help rule out infection.

## Treatment

Sarcoidosis that produces no symptoms requires no treatment. However, those severely affected with sarcoidosis require treatment with corticosteroids. Such therapy is usually continued for 1 to 2 years, but some patients may need lifelong therapy. Immunosuppressive agents, such as methotrexate, azathioprine, and cyclophosphamide, may also be used. If organ failure occurs (although this is rare), transplantation may be required. Other measures include a low-calcium diet and avoidance of direct exposure to sunlight in patients with hypercalcemia.

## Special Considerations

- ◆ Watch for and report any complications. Be aware of abnormal laboratory results (e.g., anemia) that could alter patient care.
- ◆ For the patient with arthralgia, administer analgesics as ordered. Record signs of progressive muscle weakness.
- ◆ Provide a nutritious, high-calorie diet and plenty of fluids. If the patient has hypercalcemia, suggest a low-calcium diet. Weigh the patient regularly to detect weight loss.
- ◆ Monitor respiratory function. Check chest X-rays for the extent of lung involvement; note and record any bloody sputum or increase in sputum. If the patient has pulmonary hypertension or end-stage cor pulmonale, check ABG levels, observe for arrhythmias, and administer oxygen, as needed.
- ◆ Because steroids may induce or worsen diabetes mellitus, perform fingerstick glucose tests at least every 12 hours at the beginning of steroid therapy. Also, watch for other steroid adverse effects, such as fluid retention, electrolyte imbalance (especially hypokalemia), moon face, hypertension, and personality change. During or after steroid withdrawal

(particularly in association with infection or other types of stress), watch for and report vomiting, orthostatic hypotension, hypoglycemia, restlessness, anorexia, malaise, and fatigue. Remember that the patient on long-term or high-dose steroid therapy is vulnerable to infection.

- When preparing the patient for discharge, stress the need for compliance with prescribed steroid therapy and regular, careful follow-up examinations and treatment. Refer the patient with failing vision to community support and resource groups and the American Foundation for the Blind, if necessary.

## SEVERE ACUTE RESPIRATORY SYNDROME

SARS is a viral respiratory infection that can progress to pneumonia and, eventually, death. The disease was first recognized in 2003 with outbreaks in China, Canada, Singapore, Taiwan, and Vietnam, with other countries—including the United States—reporting smaller numbers of cases.

### Causes and Incidence

SARS is caused by the SARS-associated coronavirus (SARS-CoV). Coronaviruses are a common cause of mild respiratory illnesses in humans, but researchers believe that a virus may have mutated, allowing it to cause this potentially life-threatening disease.

Close contact with a person who's infected with SARS, including contact with infectious aerosolized droplets or body secretions, is the method of transmission. Most people who contracted the disease during the 2003 outbreak contracted it during travel to endemic areas. However, the virus has been found to live on hands, tissues, and other surfaces for up to 6 hours in its droplet form. It has also been found to live in the stool of people with SARS for up to 4 days. The virus may be able to live for months or years in below-freezing temperatures.

### Pathophysiology

SARS-CoV is replicated primarily in the lungs but is also found to have the same ability in the gastrointestinal (GI) tract. Once an infection has been established, the virus will cause lysis of cells in the lungs and GI tissues and initiating an immune response. Most tissue damage takes place in the pulmonary tissues and alveoli.

### Complications

- ◆ Respiratory failure
- ◆ Liver failure
- ◆ Heart failure
- ◆ Myelodysplastic syndromes
- ◆ Death

## Signs and Symptoms

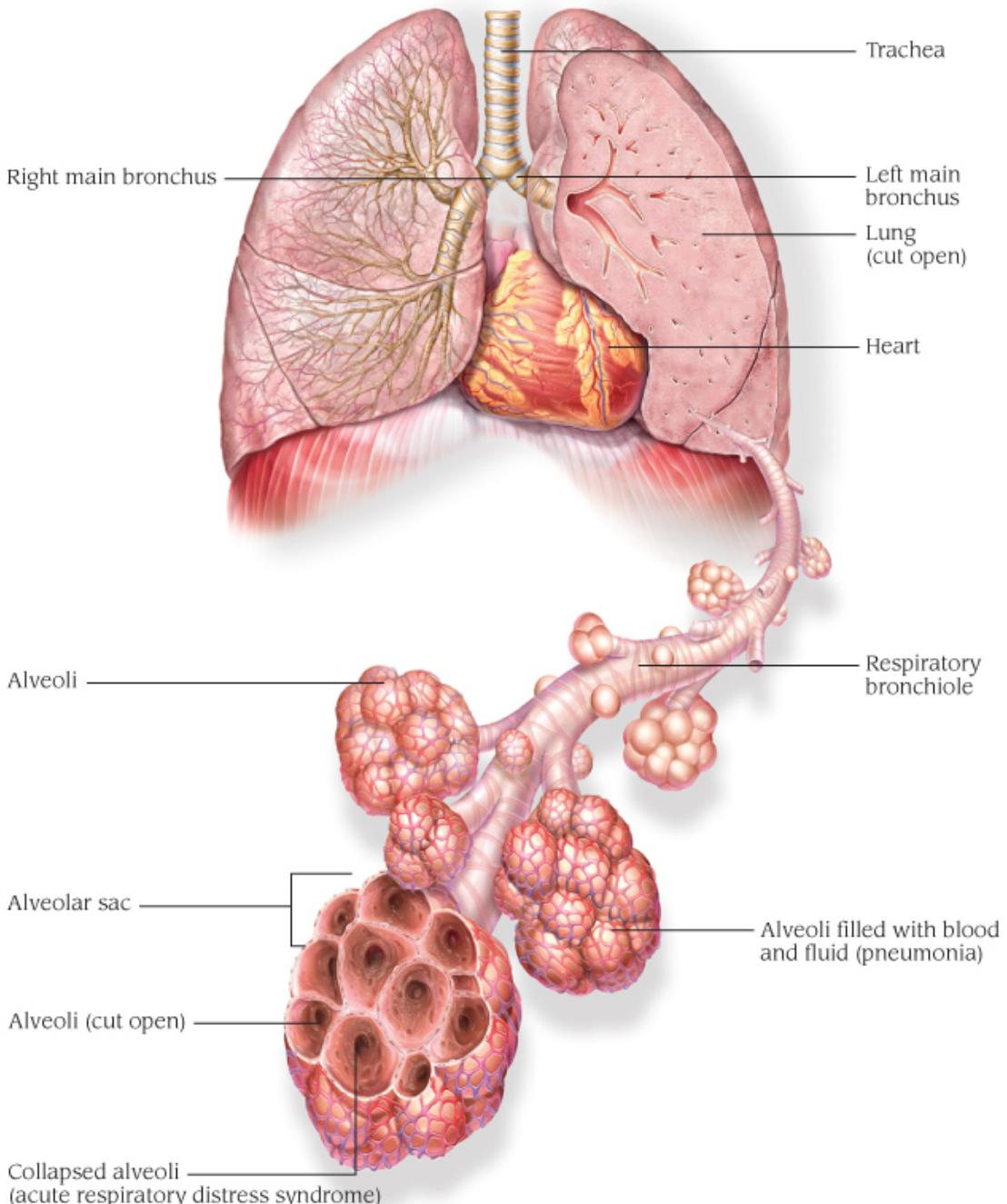
The incubation period for SARS is typically 3 to 5 days but may last as long as 14 days. Initial signs and symptoms include fever, shortness of breath and other minor respiratory symptoms, general discomfort, headache, rigors, chills, myalgia, sore throat, and dry cough. Some individuals may develop diarrhea or a rash. Later complications include respiratory failure, liver failure, heart failure, myelodysplastic syndromes, and death.

## Diagnosis

Diagnosis of severe respiratory illness is made when the patient has a fever greater than 100.4° F (38° C) or upon clinical findings of lower respiratory illness and a chest X-ray demonstrating pneumonia or ARDS. (See *Lungs and alveoli in SARS*, page 124.)

### Lungs and Alveoli in SARS

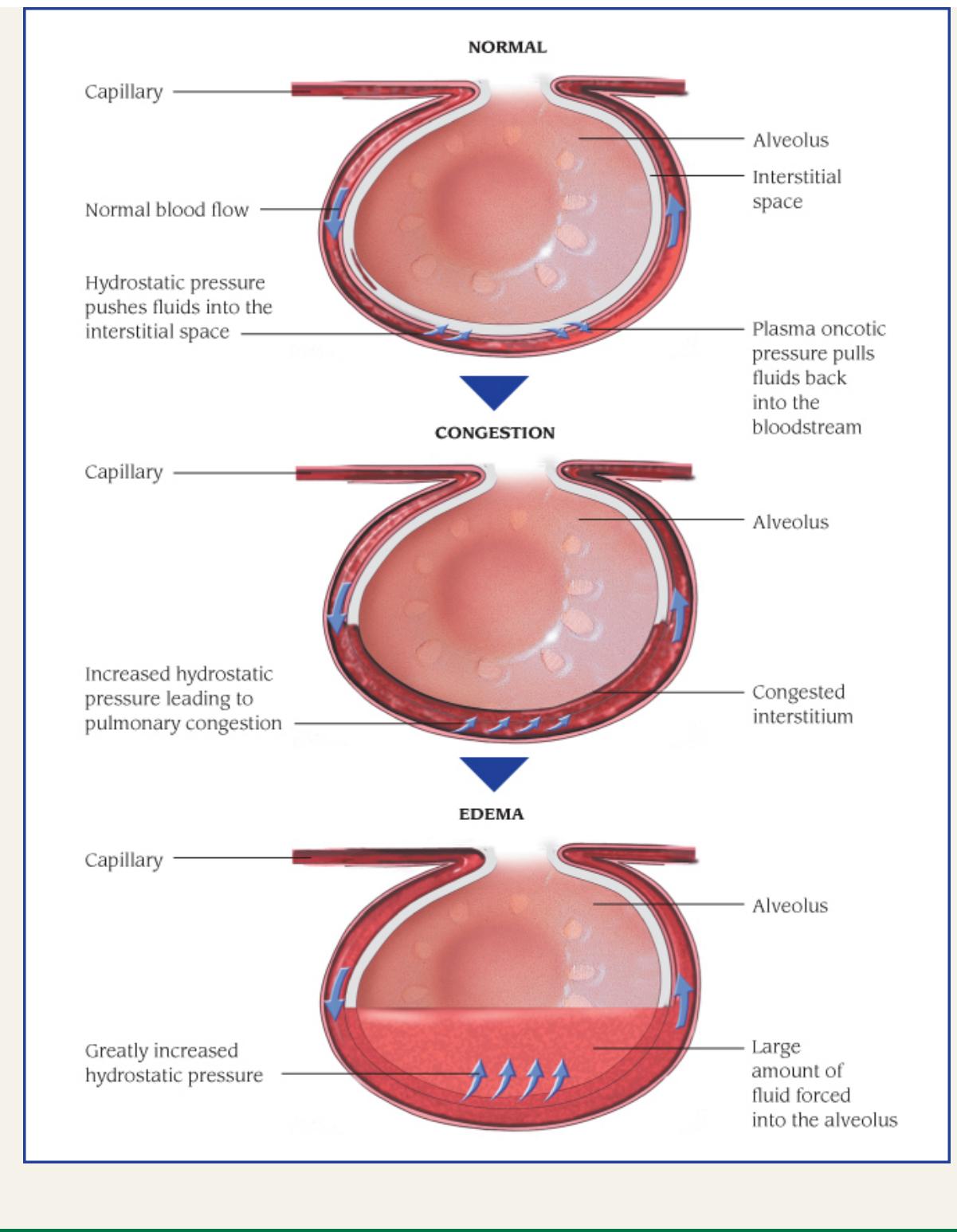
Severe acute respiratory syndrome (SARS) is a viral respiratory infection that can progress from minor respiratory symptoms to pneumonia and eventually death.



## PATHOPHYSIOLOGY HOW PULMONARY EDEMA DEVELOPS

In pulmonary edema, diminished function of the left ventricle causes blood to pool there and in the left atrium. Eventually, blood backs up into the pulmonary veins and capillaries.

Increasing capillary hydrostatic pressure pushes fluid into the interstitial spaces and alveoli. The illustrations below show a normal alveolus and the effects of pulmonary edema.



Laboratory validation for the virus includes cell culture of SARS-CoV, detection of SARS-CoV ribonucleic acid by the reverse transcription polymerase chain reaction (PCR) test, or detection of serum antibodies to

SARS-CoV. Detectable levels of antibodies may not be present until 21 days after the onset of illness, but some individuals develop antibodies within 14 days. A negative PCR, antibody test, or cell culture doesn't rule out the diagnosis.

## Treatment

Treatment is symptomatic and supportive and includes maintenance of a patent airway and adequate nutrition. Other treatment measures include supplemental oxygen, chest physiotherapy, or mechanical ventilation. In addition to standard precautions, contact precautions requiring gowns and gloves for all patient contacts and airborne precautions utilizing a negative-pressure isolation room and properly fitted N95 respirators are recommended for patients who are hospitalized. Quarantine may be used to prevent the spread of infection.

Antibiotics may be given to treat bacterial causes of atypical pneumonia. Antiviral medications have also been used. High doses of corticosteroids have been used to reduce lung inflammation. In some serious cases, serum from individuals who have already recovered from SARS (convalescent serum) has been given. The general benefit of these treatments hasn't been determined conclusively.

## Special Considerations

- ◆ Report suspected cases of SARS to local and national health organizations.
- ◆ Frequently monitor the patient's vital signs and respiratory status.
- ◆ Maintain isolation as recommended. The patient will need emotional support to deal with anxiety and fear related to the diagnosis of SARS and as a result of isolation.
- ◆ Provide patient and family teaching, including the importance of frequent handwashing, covering the mouth and nose when coughing or sneezing, and avoiding close personal contact while infected or potentially infected. Instruct the patient and family that such items as eating utensils, towels, and bedding shouldn't be shared until they have been washed with soap and hot water and that disposable gloves and household disinfectant should be used to clean any surface that may have been exposed to the patient's body fluids.
- ◆ Emphasize to the patient the importance of not going to work, school, or other public places, as recommended by the healthcare provider.

## LUNG ABSCESS

Lung abscess is a lung infection accompanied by pus accumulation and tissue destruction. The abscess may be putrid (due to anaerobic bacteria) or nonputrid (due to anaerobes or aerobes) and usually has a well-defined border. The availability of effective antibiotics has made lung abscess much less common than it was in the past.

### Causes and Incidence

Lung abscess is a manifestation of necrotizing pneumonia, generally the result of aspiration of oropharyngeal contents. Poor oral hygiene with dental or gingival (gum) disease is strongly associated with putrid lung abscess. Septic pulmonary emboli commonly produce cavitary lesions. Infected cystic lung lesions and cavitating bronchial carcinoma must be distinguished from lung abscesses.

### Pathophysiology

The development of a lung abscess is a response to the introduction of a pathogen, likely from the mouth, into the lower airways. These anaerobes are not cleared away with the body's initial immune response and create a thick-walled cavity filled with purulent material in response to the infection. They can lead to tissue necrosis or possibly the destruction of the involved lung parenchyma.

### Complications

- ◆ Rupture into pleural space, resulting in empyema
- ◆ Massive hemorrhage

### Signs and Symptoms

The clinical effects of lung abscess include a cough that may produce bloody, purulent, or foul-smelling sputum, pleuritic chest pain, dyspnea, excessive sweating, chills, fever, headache, malaise, diaphoresis, and weight loss. Chronic lung abscess may cause localized bronchiectasis. Failure of an abscess to improve with antibiotic treatment suggests possible underlying neoplasm or other causes of obstruction.

### Diagnosis

- ◆ Auscultation of the chest may reveal crackles and decreased breath sounds.

- ◆ Chest X-ray shows a localized infiltrate with one or more clear spaces, usually containing air–fluid levels.
- ◆ Percutaneous aspiration or bronchoscopy may be used to obtain cultures to identify the causative organism. Bronchoscopy is only used if abscess resolution is eventful and the patient’s condition permits it.
- ◆ Blood cultures, Gram stain, and sputum culture are also used to detect the causative organism; leukocytosis (WBC count greater than 10,000/ $\mu$ L) is commonly present.

## Treatment

Treatment consists of prolonged antibiotic therapy, commonly lasting for months, until radiographic resolution or definite stability occurs. Symptoms usually disappear in a few weeks. Postural drainage may facilitate discharge of necrotic material into the upper airways where expectoration is possible; oxygen therapy may relieve hypoxemia. Poor response to therapy requires resection of the lesion or removal of the diseased section of the lung. All patients need rigorous follow-up and serial chest X-rays.

## Special Considerations

- ◆ Help the patient with chest physiotherapy (including coughing and deep breathing), increase fluid intake to loosen secretions, and provide a quiet, restful atmosphere.
- ◆ To prevent lung abscess in the unconscious patient and the patient with seizures, first prevent aspiration of secretions. Do this by suctioning the patient and by positioning in such a way to promote drainage of secretions.

## HEMOTHORAX

In hemothorax, blood from damaged intercostal, pleural, mediastinal, and (infrequently) lung parenchymal vessels enters the pleural cavity. Depending on the amount of bleeding and the underlying cause, hemothorax may be associated with varying degrees of lung collapse and mediastinal shift. Pneumothorax—air in the pleural cavity—commonly accompanies hemothorax.

## Causes and Incidence

Hemothorax usually results from blunt or penetrating chest trauma; in fact, about 25% of patients with such trauma have hemothorax. In some cases, it

results from thoracic surgery, pulmonary infarction, neoplasm, dissecting thoracic aneurysm, or as a complication of TB or anticoagulant therapy.

## **Pathophysiology**

A hemothorax occurs when a disruption of the chest wall tissues or intrathoracic structures allows blood to enter the pleural space. Depending on the speed and volume of the circulating blood, there can be a significant hemodynamic shift including the early stages of shock.

## **Complications**

- ◆ Mediastinal shift
- ◆ Ventilatory compromise
- ◆ Lung collapse
- ◆ Cardiopulmonary arrest

## **Signs and Symptoms**

The patient with hemothorax may experience chest pain, tachypnea, and mild to severe dyspnea, depending on the amount of blood in the pleural cavity and associated pathologic conditions. If respiratory failure results, the patient may appear anxious, restless, possibly stuporous, and cyanotic; marked blood loss produces hypotension and shock. The affected side of the chest expands and stiffens, whereas the unaffected side rises and falls with the patient's breaths.

## **Diagnosis**

Characteristic clinical signs and a history of trauma strongly suggest hemothorax. Percussion and auscultation reveal dullness and decreased to absent breath sounds over the affected side. Thoracentesis yields blood or serosanguineous fluid; chest X-rays show pleural fluid with or without mediastinal shift. ABG analysis may reveal respiratory failure; Hb may be decreased, depending on the amount of blood lost.

## **Treatment**

Treatment is designed to stabilize the patient's condition, stop the bleeding, evacuate blood from the pleural space, and re-expand the underlying lung. Mild hemothorax usually clears in 10 to 14 days, requiring only observation for further bleeding. In severe hemothorax, thoracentesis not only serves as a diagnostic tool, but also removes fluid from the pleural cavity.

After the diagnosis is confirmed, a chest tube is inserted into the sixth intercostal space at the posterior axillary line. Suction may be used; a large-bore tube is used to prevent clot blockage. If the chest tube doesn't improve the patient's condition, they may need a thoracotomy to evacuate blood and clots and to control bleeding.

## **Special Considerations**

- ◆ Give oxygen by face mask or nasal cannula.
- ◆ Give I.V. fluids and blood transfusions, as ordered, to treat shock. Monitor pulse oximetry and ABG levels often.
- ◆ Explain all procedures to the patient to allay their fears. Assist with thoracentesis. Warn the patient not to cough during this procedure.
- ◆ Carefully observe chest tube drainage and record the volume drained (at least every hour). Milk the chest tube (only if necessary and according to facility and physician protocols) to keep it open and free from clots. If the tube is warm and full of blood and the bloody fluid level in the water-seal bottle is rising rapidly, report this at once. The patient may need immediate surgery.
- ◆ Watch the patient closely for pallor and gasping respirations. Monitor vital signs diligently. Falling blood pressure, rising pulse rate, and rising respiratory rate may indicate shock or massive bleeding.

## **PULMONARY HYPERTENSION**

Pulmonary hypertension occurs when PAP rises above normal for reasons other than aging or altitude. No definitive set of values is used to diagnose pulmonary hypertension, but the National Institutes of Health requires a mean PAP of 25 mm Hg or more. The prognosis depends on the cause of the underlying disorder, but the long-term prognosis is poor. Within 5 years of diagnosis, only 25% of patients are still alive.

### **Causes and Incidence**

Pulmonary hypertension begins as hypertrophy of the small pulmonary arteries. The medial and intimal muscle layers of these vessels thicken, decreasing distensibility and increasing resistance. This disorder then progresses to vascular sclerosis and obliteration of small vessels.

In most cases, pulmonary hypertension occurs secondary to an underlying disease process, including:

- ◆ *alveolar hypoventilation* from COPD (most common cause in the United States), sarcoidosis, diffuse interstitial disease, pulmonary metastasis, and certain diseases such as scleroderma. (In these disorders, pulmonary vascular resistance occurs secondary to hypoxemia and destruction of the alveolocapillary bed. Other disorders that cause alveolar hypoventilation without lung tissue damage include obesity, kyphoscoliosis, and obstructive sleep apnea.)
- ◆ *vascular obstruction* from pulmonary embolism, vasculitis, and disorders that cause obstruction of small or large pulmonary veins, such as left atrial myxoma, idiopathic venoocclusive disease, fibrosing mediastinitis, and mediastinal neoplasm
- ◆ *primary cardiac disease*, which may be congenital or acquired. Congenital defects that cause left-to-right shunting of blood—such as patent ductus arteriosus or atrial or ventricular septal defect—increase blood flow into the lungs and, consequently, raise pulmonary vascular pressure. Acquired cardiac diseases, such as rheumatic valvular disease and mitral stenosis, increase pulmonary venous pressure by restricting blood flow returning to the heart

Primary (or idiopathic) pulmonary hypertension is rare, occurring most commonly—and with no known cause—in women between 20 and 40 years old. Secondary pulmonary hypertension results from existing cardiac, pulmonary, thromboembolic, or collagen vascular diseases or from the use of certain drugs.

## **Pathophysiology**

The primary pathogenic mechanism involved in pulmonary hypertension is vascular resistance. This resistance typically results from vasoconstriction, remodeling, or a formation of a microthrombus in the pulmonary arteries or arterioles.

## **Complications**

- ◆ Cor pulmonale
- ◆ Cardiac failure
- ◆ Cardiac arrest

## **Signs and Symptoms**

Most patients complain of increasing dyspnea on exertion, weakness, syncope, and fatigability. Many also show signs of right-sided heart failure, including peripheral edema, ascites, jugular vein distention, and hepatomegaly. Other clinical effects vary with the underlying disorder.

## Diagnosis

Characteristic diagnostic findings include:

- ◆ Auscultation reveals abnormalities associated with the underlying disorder.
- ◆ ABG analysis indicates hypoxemia (decreased Pao<sub>2</sub>).
- ◆ Electrocardiography shows right axis deviation and tall or peaked P waves in inferior leads in the patient with right ventricular hypertrophy.
- ◆ Cardiac catheterization reveals pulmonary systolic pressure above 30 mm Hg as well as increased PAWP if the underlying cause is left atrial myxoma, mitral stenosis, or left-sided heart failure (otherwise normal).
- ◆ Pulmonary angiography detects filling defects in pulmonary vasculature such as those that develop in patients with pulmonary emboli.
- ◆ PFTs may show decreased flow rates and increased residual volume in underlying obstructive disease and decreased TLC in underlying restrictive disease.

## Treatment

Treatment usually includes oxygen therapy to decrease hypoxemia and resulting pulmonary vascular resistance. It may also include vasodilator therapy (nifedipine [Procardia], diltiazem [Cardizem], or prostaglandin E). For patients with right-sided heart failure, treatment also includes fluid restriction, cardiac glycosides to increase cardiac output, and diuretics to decrease intravascular volume and extravascular fluid accumulation. Treatment also aims to correct the underlying cause.

Some patients with pulmonary hypertension may be candidates for heart-lung transplantation to improve their chances of survival.

## Special Considerations

Pulmonary hypertension requires keen observation and careful monitoring as well as skilled supportive care.

- ◆ Administer oxygen therapy as ordered and observe the patient's response. Report any signs of increasing dyspnea to the physician so they can adjust treatment accordingly.

- ◆ Monitor ABG levels for acidosis and hypoxemia. Report any change in the patient's level of consciousness at once.
- ◆ When caring for a patient with right-sided heart failure, especially one receiving diuretics, record their weight daily, carefully measure intake and output, and explain all medications and diet restrictions. Check for worsening jugular vein distention, which may indicate fluid overload.
- ◆ Monitor the patient's vital signs, especially blood pressure and heart rate. Watch for hypotension and tachycardia. If they have a pulmonary artery catheter, check PAP and PAWP, as indicated. Report any changes.
- ◆ Before discharge, help the patient adjust to the limitations imposed by this disorder. Advise against overexertion and suggest frequent rest periods between activities. Refer the patient to the social services department if they will need special equipment, such as oxygen equipment, for home use. Make sure that they understand the prescribed medications and diet and the need to weigh themselves daily.

## **PLEURAL EFFUSION AND EMPYEMA**

Pleural effusion is an excess of fluid in the pleural space. Normally, this space contains a small amount of extracellular fluid that lubricates the pleural surfaces. Increased production or inadequate removal of this fluid results in pleural effusion. Empyema is the accumulation of pus and necrotic tissue in the pleural space. Blood (hemothorax) and chyle (chylothorax) may also collect in this space.

### **Causes and Incidence**

The balance of osmotic and hydrostatic pressures in parietal pleural capillaries normally results in fluid movement into the pleural space. Balanced pressures in visceral pleural capillaries promote reabsorption of this fluid. Effusions frequently result from heart failure, hepatic disease with ascites, peritoneal dialysis, hypoalbuminemia, and disorders resulting in overexpanded intravascular volume.

Exudative pleural effusions occur with TB, subphrenic abscess, pancreatitis, bacterial or fungal pneumonitis or empyema, malignancy, pulmonary embolism with or without infarction, collagen disease (LE and rheumatoid arthritis), myxedema, and chest trauma.

Empyema is usually associated with infection in the pleural space. Such infection may be idiopathic or may be related to pneumonitis, carcinoma, perforation, or esophageal rupture.

## **Pathophysiology**

Pleural effusions result from excessive hydrostatic pressure or decreased osmotic pressure causing excessive amounts of fluid to pass across intact capillaries. The result is a transudative pleural effusion, an ultrafiltrate of plasma containing low concentrations of protein. Exudative pleural effusions result when capillaries exhibit increased permeability with or without changes in hydrostatic and colloid osmotic pressures, allowing protein-rich fluid to leak into the pleural space. An empyema results from an infection in the pleural space or accumulated fluid.

## **Complications**

- ◆ Atelectasis
- ◆ Infection
- ◆ Hypoxemia

## **Signs and Symptoms**

Patients with pleural effusion characteristically display symptoms relating to the underlying pathologic condition. Most patients with large effusions, particularly those with underlying pulmonary disease, complain of dyspnea. Those with effusions associated with pleurisy complain of pleuritic chest pain. Other clinical features depend on the cause of the effusion. Patients with empyema also develop fever and malaise.

## **Diagnosis**

Auscultation of the chest reveals decreased breath sounds; percussion detects dullness over the effused area, which doesn't change with breathing. Chest X-ray shows fluid in dependent regions. However, diagnosis also requires other tests to distinguish transudative from exudative effusions and to help pinpoint the underlying disorder.

The most useful test is thoracentesis, in which pleural fluid is analyzed in the laboratory to show components. Acute inflammatory WBCs and microorganisms may be evident in empyema.

In addition, if a pleural effusion results from esophageal rupture or pancreatitis, fluid amylase levels are usually higher than serum levels. Aspirated fluid may be tested for LE cells, antinuclear antibodies, and neoplastic cells. It may also be analyzed for color and consistency; acid-fast bacillus (AFB), fungal, and bacterial cultures; and triglycerides (in

chylothorax). Cell analysis shows leukocytosis in empyema. A negative tuberculin skin test strongly rules against TB as the cause. In exudative pleural effusions in which thoracentesis isn't definitive, pleural biopsy may be done. This is particularly useful for confirming TB or malignancy.

## Treatment

Depending on the amount of fluid present, symptomatic effusion may require thoracentesis to remove fluid or careful monitoring of the patient's own reabsorption of the fluid. Hemothorax requires drainage to prevent fibrothorax formation. Pleural effusions associated with lung cancer commonly reaccumulate quickly. If a chest tube is inserted to drain the fluid, a sclerosing agent, such as talc, may be injected through the tube to cause adhesions between the parietal and visceral pleura, thereby obliterating the potential space for fluid to recollect.

Treatment of empyema requires insertion of one or more chest tubes after thoracentesis, to allow drainage of purulent material, and possibly decortication (surgical removal of the thick coating over the lung) or rib resection to allow open drainage and lung expansion. Empyema also requires parenteral antibiotics. Associated hypoxia requires oxygen administration.

## Special Considerations

- ◆ Explain thoracentesis to the patient. Before the procedure, tell the patient to expect a stinging sensation from the local anesthetic and a feeling of pressure when the needle is inserted. Instruct them to tell you immediately if they feel uncomfortable or has difficulty breathing during the procedure.
- ◆ Reassure the patient during thoracentesis. Remind them to breathe normally and avoid sudden movements, such as coughing or sighing. Monitor vital signs and watch for syncope. If fluid is removed too quickly, the patient may suffer bradycardia, hypotension, pain, pulmonary edema, or even cardiac arrest. Watch for respiratory distress or pneumothorax (sudden onset of dyspnea and cyanosis) after thoracentesis.
- ◆ Administer oxygen and, in empyema, antibiotics, as ordered.
- ◆ Encourage the patient to perform deep-breathing exercises to promote lung expansion. Use an incentive spirometer to promote deep breathing.
- ◆ Provide meticulous chest tube care, and use sterile technique for changing dressings around the tube insertion site in empyema. Ensure tube patency by watching for fluctuations of fluid or air bubbling in the underwater seal

chamber. Continuous bubbling may indicate an air leak. Record the amount, color, and consistency of any tube drainage.

- ◆ If the patient has open drainage through a rib resection or intercostal tube, use hand and dressing precautions. Because weeks of such drainage are usually necessary to obliterate the space, make visiting nurse referrals for the patient who will be discharged with the tube in place.
- ◆ If pleural effusion was a complication of pneumonia or influenza, advise prompt medical attention for upper respiratory infections.

## PLEURISY

Pleurisy, also known as *pleuritis*, is an inflammation of the visceral and parietal pleurae that line the inside of the thoracic cage and envelop the lungs.

### Causes and Incidence

Pleurisy develops as a complication of pneumonia, TB, viruses, systemic LE, rheumatoid arthritis, uremia, Dressler syndrome, certain cancers, pulmonary infarction, and chest trauma. Pleuritic pain is caused by the inflammation or irritation of sensory nerve endings in the parietal pleura. As the lungs inflate and deflate, the visceral pleura covering the lungs moves against the fixed parietal pleura lining the pleural space, causing pain. This disorder usually begins suddenly.

In the United States, pleural effusions develop in 36% to 66% of hospitalized patients with bacterial pneumonia.

### Pathophysiology

There are pain fibers located in the two layers of the pleura; the visceral pleural covers the lung and the parietal pleural line the inner chest wall. Pleurisy or pleuritis is the result of the inflammation of these pleuritic layers, usually from an infectious source.

### Complications

- ◆ Pneumonia
- ◆ Pleural effusion
- ◆ Lung collapse

### Signs and Symptoms

Sharp, stabbing pain that increases with deep breathing may be so severe that it limits movement on the affected side. Dyspnea also occurs. Other symptoms vary according to the underlying pathologic process.

## Diagnosis

Auscultation of the chest reveals a characteristic pleural friction rub—a coarse, creaky sound heard during late inspiration and early expiration, directly over the area of pleural inflammation. Palpation over the affected area may reveal coarse vibration. Chest X-ray, ultrasound of the chest, and thoracentesis may aid in diagnosis.

## Treatment

Treatment is directed at the underlying cause; bacterial infections are treated with appropriate antibiotics, TB requires special treatment, and viral infections may be permitted to run their course. Treatment also includes measures to relieve symptoms, such as anti-inflammatory agents, analgesics, and bed rest. Severe pain may require an intercostal nerve block of two or three intercostal nerves. Pleurisy with pleural effusion calls for thoracentesis as a therapeutic and diagnostic measure.

## Special Considerations

- ◆ Stress the importance of bed rest and plan your care to allow the patient as much uninterrupted rest as possible.
- ◆ Administer antitussives and pain medication, as ordered, but be careful not to overmedicate. If the pain requires an opioid analgesic, warn the patient who's about to be discharged to avoid overuse because such medication depresses coughing and respiration.
- ◆ Encourage the patient to cough. Tell them to apply firm pressure at the site of the pain during coughing exercises to minimize pain.

## Chronic Disorders

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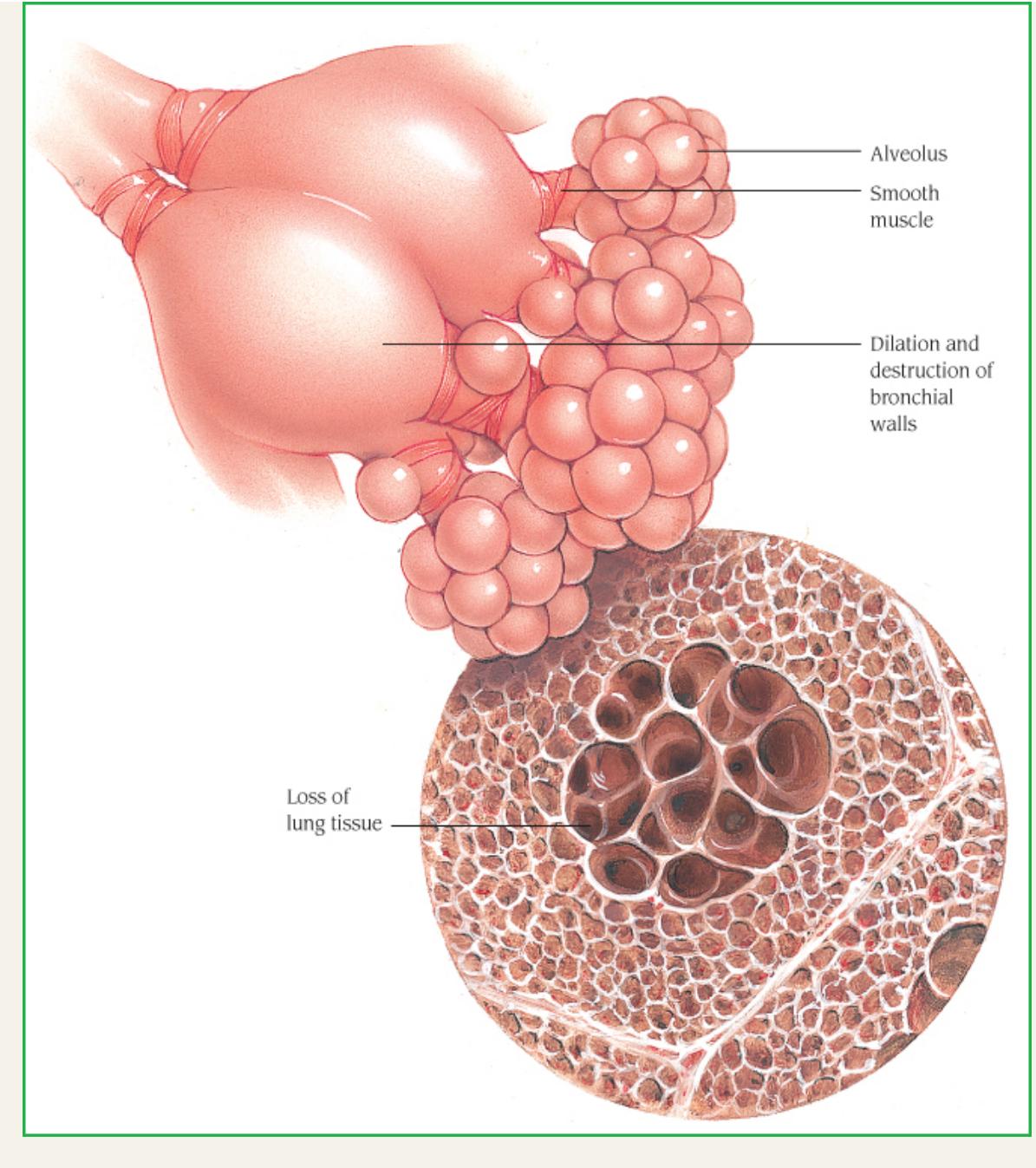
### CHRONIC OBSTRUCTIVE PULMONARY DISEASE

COPD is chronic airway obstruction that results from emphysema, chronic bronchitis, asthma, or any combination of these disorders. (See *How pulmonary edema develops*, page 131. Also see *Lung changes in emphysema*,

page 132.) Usually, more than one of these underlying conditions coexist; in most cases, bronchitis and emphysema occur together. It doesn't always produce symptoms and causes only minimal disability in many patients. However, COPD tends to worsen with time.

## **Lung Changes in Emphysema**

A form of chronic obstructive pulmonary disease, emphysema is the abnormal, permanent enlargement of the acini accompanied by the destruction of the alveolar walls. Obstruction results from tissue changes, rather than mucus production, as occurs in asthma and chronic bronchitis. The distinguishing characteristic of emphysema is airflow limitation caused by a lack of elastic recoil in the lungs.



## Causes and Incidence

Predisposing factors include cigarette smoking, recurrent or chronic respiratory infections, air pollution, occupational exposure to chemicals, and allergies. Early inflammatory changes may reverse if the patient stops smoking before lung destruction is extensive. Familial and hereditary factors

(such as deficiency of alpha<sub>1</sub>-antitrypsin) may also predispose a person to COPD.

The most common chronic lung disease, COPD (also known as *chronic obstructive lung disease*) affects an estimated 17 million Americans, and its incidence is rising. It affects more males than females, probably because until recently men were more likely to smoke heavily. COPD occurs mostly in people older than age 40.

## Pathophysiology

Repeated exposure to carcinogens, such as smoking, is by far the most important causative action—it impairs ciliary action and macrophage function, inflames airways, increases mucus production, destroys alveolar septae, and causes peribronchiolar fibrosis.

## Complications

- ◆ Overwhelming disability
- ◆ Cor pulmonale
- ◆ Severe respiratory failure
- ◆ Death

## Signs and Symptoms

The typical patient, a long-term cigarette smoker, has no symptoms until middle age. The ability to exercise or do strenuous work gradually starts to decline, and the patient begins to develop a productive cough. These signs are subtle at first but become more pronounced as the patient gets older and the disease progresses. Eventually the patient may develop dyspnea on minimal exertion, frequent respiratory infections, intermittent or continuous hypoxemia, and grossly abnormal pulmonary function studies. Advanced COPD may cause severe dyspnea, overwhelming disability, cor pulmonale, severe respiratory failure, and death.

## Diagnosis

For specific diagnostic tests used to determine COPD, see *Types of chronic obstructive pulmonary disease*.

## Treatment

Treatment is designed to relieve symptoms and prevent complications. Because most patients with COPD receive outpatient treatment, they need comprehensive teaching to help them comply with therapy and understand the nature of this chronic, progressive disease. If programs in pulmonary rehabilitation are available, encourage patient to enroll.

Urge the patient to stop smoking. Provide smoking cessation counseling or refer them to a program. Avoid other respiratory irritants, such as secondhand smoke, aerosol spray products, and outdoor air pollution. An air conditioner with an air filter in the home may be helpful.

The patient is usually treated with beta-agonist bronchodilators (albuterol or salmeterol), anticholinergic bronchodilators (ipratropium), and corticosteroids (beclomethasone or triamcinolone). These are usually given by metered-dose inhaler, requiring that the patient be taught the correct administration technique.

Antibiotics are used to treat respiratory infections. Stress the need to complete the prescribed course of antibiotic therapy.

## **Special Considerations**

- ◆ Teach the patient and family how to recognize early signs of infection; warn the patient to avoid contact with people with respiratory infections. Encourage good oral hygiene to help prevent infection. Pneumococcal vaccination and annual influenza vaccinations are important preventive measures.
- ◆ To promote ventilation and reduce air trapping, teach the patient to breathe slowly, prolong expirations to two to three times the duration of inspiration, and to exhale through pursed lips.
- ◆ To help mobilize secretions, teach the patient how to cough effectively. If the patient with copious secretions has difficulty mobilizing secretions, teach his or her family how to perform postural drainage and chest physiotherapy. If secretions are thick, urge the patient to drink 12 to 15 glasses of fluid per day. A home humidifier may be beneficial, particularly in the winter.
- ◆ Administer low concentrations of oxygen as ordered. Perform blood gas analysis to determine the patient's oxygen needs and to avoid CO<sub>2</sub> narcosis. If the patient is to continue oxygen therapy at home, teach them how to use the equipment correctly. The patient with COPD rarely requires more than 2 to 3 L/minute to maintain adequate oxygenation. Higher flow

rates will further increase the  $\text{Pao}_2$ , but the patient whose ventilatory drive is largely based on hypoxemia commonly develops markedly increased  $\text{Paco}_2$ . In these cases, chemoreceptors in the brain are relatively insensitive to the increase in  $\text{CO}_2$ . Teach the patient and family that excessive oxygen therapy may eliminate the hypoxic respiratory drive, causing confusion and drowsiness, signs of  $\text{CO}_2$  narcosis.

- ◆ Emphasize the importance of a balanced diet. Because the patient may tire easily when eating, suggest that they eat frequent, small meals and consider using oxygen, administered by nasal cannula, during meals.
- ◆ Help the patient and family adjust their lifestyles to accommodate the limitations imposed by this debilitating chronic disease. Instruct the patient to allow for daily rest periods and to exercise daily as the provider directs.
- ◆ As COPD progresses, encourage the patient to discuss their fears.
- ◆ To help prevent COPD, advise all patients, especially those with a family history of COPD or those in its early stages, not to smoke.
- ◆ Assist in the early detection of COPD by urging persons to have periodic physical examinations, including spirometry and medical evaluation of a chronic cough, and to seek treatment for recurring respiratory infections promptly.
- ◆ Lung volume reduction surgery is a new procedure for carefully selected patients with primarily emphysema. Nonfunctional parts of the lung (tissue filled with disease and providing little ventilation or perfusion) are surgically removed. Removal allows more functional lung tissue to expand and the diaphragm to return to its normally elevated position.

## **BRONCHIECTASIS**

A condition marked by chronic abnormal dilation of bronchi and destruction of bronchial walls, bronchiectasis can occur throughout the tracheobronchial tree or can be confined to one segment or lobe. However, it's usually bilateral and involves the basilar segments of the lower lobes. This disease has three forms: cylindrical (fusiform), varicose, and saccular (cystic). Bronchiectasis is irreversible once established.

### **Causes and Incidence**

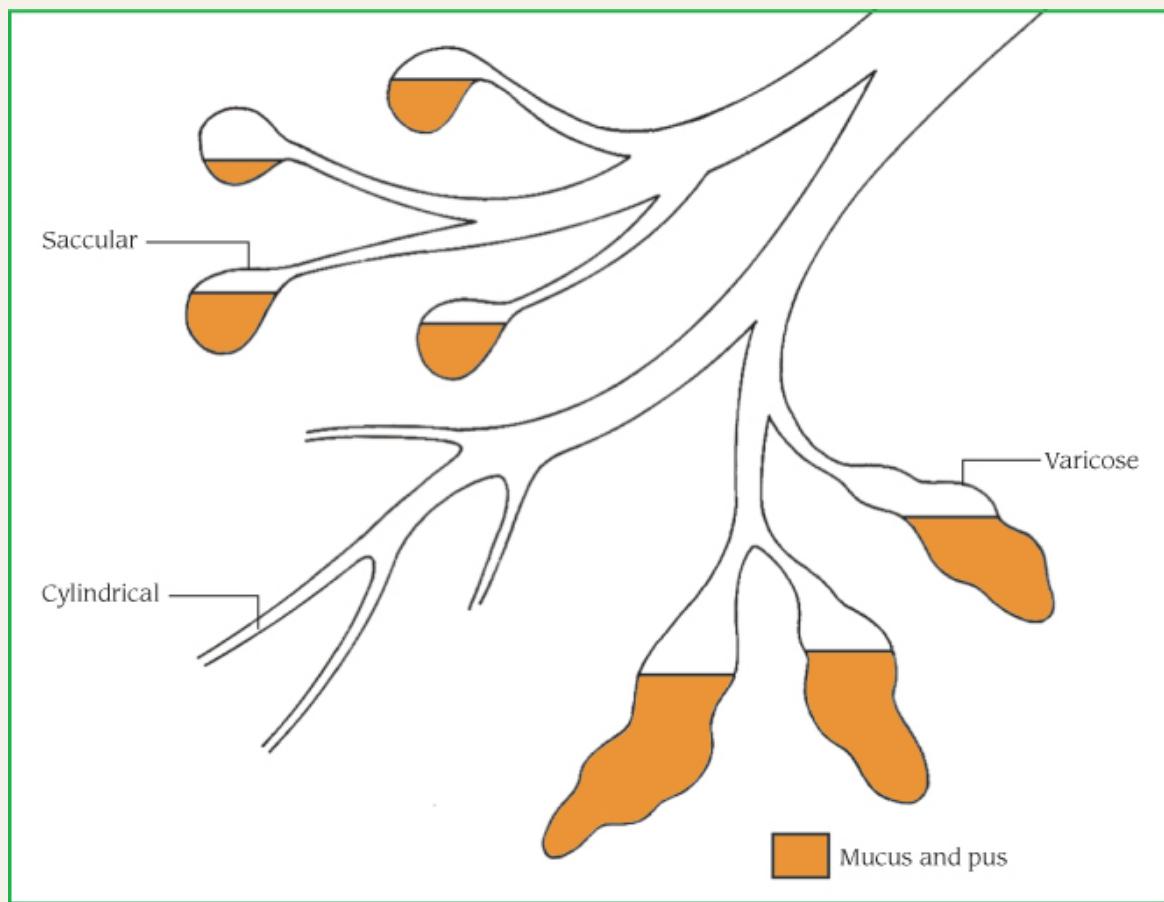
Because of the availability of antibiotics to treat acute respiratory tract infections, the incidence of bronchiectasis has dramatically decreased in the

past 20 years. Incidence is highest among Eskimos and the Maoris of New Zealand. It affects people of both sexes and all ages.

The different forms of bronchiectasis may occur separately or simultaneously. In *cylindrical bronchiectasis*, the bronchi expand unevenly, with little change in diameter, and end suddenly in a squared-off fashion. In *varicose bronchiectasis*, abnormal, irregular dilation and narrowing of the bronchi give the appearance of varicose veins. In *saccular bronchiectasis*, many large dilations end in sacs. These sacs balloon into pus-filled cavities as they approach the periphery and are then called saccules. (See *Forms of bronchial dilatation*, page 134.)

## Forms of Bronchial Dilatation

Dilatations of the air sacs occur because of bronchiectasis, as depicted below.



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This disease results from conditions associated with repeated damage to bronchial walls and abnormal mucociliary clearance, which cause a breakdown of supporting tissue adjacent to airways. Such conditions include:

- ◆ cystic fibrosis
- ◆ immunologic disorders (e.g., agammaglobulinemia)
- ◆ recurrent, inadequately treated bacterial respiratory tract infections, such as TB, and complications of measles, pneumonia, pertussis, or influenza
- ◆ obstruction (by a foreign body [most common in children], tumor, or stenosis) in association with recurrent infection
- ◆ inhalation of corrosive gas or repeated aspiration of gastric juices into the lungs
- ◆ congenital anomalies (uncommon), such as bronchomalacia, congenital bronchiectasis, immotile cilia syndrome, and Kartagener syndrome, a variant of immotile cilia syndrome characterized by situs inversus, bronchiectasis, and either nasal polyps or sinusitis

## Pathophysiology

In bronchiectasis, hyperplastic squamous epithelium denuded of cilia replaces ulcerated columnar epithelium. Abscess formation involving all layers of the bronchial wall produces inflammatory cells and fibrous tissue, resulting in dilation and narrowing of the airways. Mucus plugs or fibrous tissue obliterates smaller bronchioles, whereas peribronchial lymphoid tissue becomes hyperplastic. Extensive vascular proliferation of bronchial circulation occurs and produces frequent hemoptysis.

## Complications

- ◆ Chronic malnutrition
- ◆ Amyloidosis
- ◆ Right ventricular failure
- ◆ Cor pulmonale

## Signs and Symptoms

Initially, bronchiectasis may be asymptomatic. When symptoms do arise, they're commonly attributed to other illnesses. The patient usually complains of frequent bouts of pneumonia or hemoptysis. The classic symptom, however, is a chronic cough that produces foul-smelling, mucopurulent

secretions in amounts ranging from less than 10 mL/day to more than 150 mL/day.

Cough and sputum production are observed in greater than 90% of bronchiectasis patients. Characteristic findings include coarse crackles during inspiration over involved lobes or segments, occasional wheezing, dyspnea, sinusitis, weight loss, anemia, malaise, clubbing, recurrent fever, chills, and other signs of infection.

Advanced bronchiectasis may produce chronic malnutrition as well as right-sided heart failure and cor pulmonale because of hypoxic pulmonary vasoconstriction.

## Diagnosis

A history of recurrent bronchial infections, pneumonia, and hemoptysis in a patient whose chest X-rays show peribronchial thickening, areas of atelectasis, and scattered cystic changes suggest bronchiectasis.

In recent years, CT scanning has supplanted bronchography as the most useful diagnostic test for bronchiectasis. It's sometimes used with high-resolution techniques to better determine anatomic changes. Bronchoscopy doesn't establish the diagnosis of bronchiectasis, but it does help to identify the source of secretions. Bronchoscopy can also be instrumental in pinpointing the site of bleeding in hemoptysis.

Other helpful laboratory tests include:

- ◆ sputum culture and Gram stain to identify predominant organisms
- ◆ complete blood count to detect anemia and leukocytosis
- ◆ PFTs to detect decreased vital capacity, expiratory flow rate, and hypoxemia. These tests also help determine the physiologic severity of the disease and the effects of therapy and help evaluate patients for surgery

When cystic fibrosis is suspected as the underlying cause of bronchiectasis, a sweat electrolyte test is useful.

## Treatment

Treatment includes antibiotics, given orally or I.V., for 7 to 10 days or until sputum production decreases. Bronchodilators, combined with postural drainage and chest percussion, help remove secretions if the patient has bronchospasm and thick, tenacious sputum. Bronchoscopy may be used to remove obstruction and secretions. Hypoxia requires oxygen therapy; severe hemoptysis commonly requires lobectomy, segmental resection, or bronchial

artery embolization if pulmonary function is poor. Long-term antibiotic therapy isn't appropriate because it may predispose the patient to serious gram-negative infections and resistant organisms.

## Special Considerations

- ◆ Provide supportive care and help the patient adjust to the permanent changes in lifestyle that irreversible lung damage necessitates. Thorough teaching is vital.
- ◆ Administer antibiotics as ordered and explain all diagnostic tests. Perform chest physiotherapy, including postural drainage and chest percussion designed for involved lobes, several times a day. The best times to do this are early morning and just before bedtime. Instruct the patient to maintain each position for 10 minutes, and then perform percussion and tell them to cough. Show family members how to perform postural drainage and percussion. Also teach the patient coughing and deep-breathing techniques to promote good ventilation and the removal of secretions.
- ◆ Advise the patient to stop smoking, if appropriate, to avoid stimulating secretions and irritating the airways. Refer them to a local self-help group.
- ◆ Provide a warm, quiet, comfortable environment, and urge the patient to rest as much as possible. Encourage balanced, high-protein meals to promote good health and tissue healing and plenty of fluids (2 to 3 qt [2 to 3 L])/day to hydrate and thin bronchial secretions. Give frequent mouth care to remove foul-smelling sputum. Teach the patient to dispose of all secretions properly. Instruct them to seek prompt attention for respiratory infections.
- ◆ Tell the patient to avoid air pollutants and people with upper respiratory tract infections. Instruct them to take medications (especially antibiotics) exactly as prescribed.



### PREVENTION

- ◆ *Treat bacterial pneumonia vigorously.*
- ◆ *Stress the need for immunization to prevent childhood diseases.*

## IDIOPATHIC PULMONARY FIBROSIS

Idiopathic pulmonary fibrosis (IPF) is a chronic and usually fatal interstitial pulmonary disease. About 50% of patients with IPF die within 5 years of

diagnosis. Once thought to be a rare condition, it's now diagnosed with much greater frequency. IPF has been known by several other names over the years, including cryptogenic fibrosing alveolitis, diffuse interstitial fibrosis, idiopathic interstitial pneumonitis, and Hamman–Rich syndrome.

## Causes and Incidence

IPF is the result of a cascade of events that involve inflammatory, immune, and fibrotic processes in the lung. However, despite many studies and hypotheses, the stimulus that begins the progression remains unknown. Speculation has revolved around viral and genetic causes, but no good evidence has been found to support either theory. However, it's clear that chronic inflammation plays an important role. Inflammation develops the injury and the fibrosis that ultimately distorts and impairs the structure and function of the alveolocapillary gas exchange surface.

IPF is slightly more common in men than in women and is more common in smokers than in nonsmokers. It usually affects people 50 to 70 years old.

## Pathophysiology

IPF pathophysiology is thought to be initiated when alveolar epithelial cells signal an injury and then activates the excessive formation of fibroblast migration and differentiation. Scarring to the lung structures occurs when the fibroblasts and myofibroblasts secrete large amounts of extracellular matrix proteins, such as collagens, decreasing the overall lung volume.

## Complications

- ◆ Respiratory failure
- ◆ Pneumonia
- ◆ Hypoxemia
- ◆ Pneumothorax
- ◆ Pulmonary hypertension

## Signs and Symptoms

The usual presenting symptoms of IPF are dyspnea and a dry, hacking, and typically paroxysmal cough. Most patients have had these symptoms for several months to 2 years before seeking medical help. Expiratory crackles, especially in the bases of the lungs, are usually heard early in the disease. Bronchial breath sounds appear later, when airway consolidation develops.

Rapid, shallow breathing occurs, especially with exertion, and clubbing has been noted in more than 40% of patients. Late in the disease, cyanosis and evidence of pulmonary hypertension (augmented S<sub>2</sub> and S<sub>3</sub> gallop) commonly occur. As the disease progresses, profound hypoxemia and severe, debilitating dyspnea are the hallmark signs.

## Diagnosis

Diagnosis begins with a thorough patient history to exclude a more common cause of interstitial lung disease.

 **CONFIRMING DIAGNOSIS** *Lung biopsy is helpful in the diagnosis of IPF. In the past, an open lung biopsy was the only acceptable procedure, but now biopsies may be done through a thoracoscope or bronchoscope.*

Histologic features of the biopsy tissue vary, depending on the stage of the disease and other factors that aren't yet completely understood. The alveolar walls are swollen with chronic inflammatory cellular infiltrate composed of mononuclear cells and polymorphonuclear leukocytes. Intra-alveolar inflammatory cells may be found in early stages. As the disease progresses, excessive collagen and fibroblasts fill the interstitium. In advanced stages, alveolar walls are destroyed and are replaced by honeycombing cysts.

Chest X-rays may show one of four distinct patterns: interstitial, reticulonodular, ground-glass, or honeycomb. Although chest X-rays are helpful in identifying the presence of an abnormality, they don't correlate well with histologic findings or PFTs in determining the severity of the disease. They also don't help distinguish inflammation from fibrosis. However, serial X-rays may help track the progression of the disease.

High-resolution CT scans provide superior views of the four patterns seen on routine X-ray film and are used routinely to help establish the diagnosis of IPF. Research is currently underway to determine whether the four patterns of abnormality seen on these scans correlate with responsiveness to treatment.

PFTs show reductions in vital capacity and TLC and impaired diffusing capacity for carbon monoxide. ABG analysis and pulse oximetry reveal hypoxemia, which may be mild when the patient is at rest early in the disease but may become severe later in the disease. Oxygenation will always deteriorate, usually to a severe level, with exertion. Serial PFTs (especially carbon monoxide diffusing capacity) and ABG values may help track the course of the disease and the patient's response to treatment.

## Treatment

Although it can't change the pathophysiology of IPF, oxygen therapy can prevent the problems related to dyspnea and tissue hypoxia in the early stages of the disease process. The patient may require little or no supplemental oxygen while at rest initially, but he or she will need more as the disease progresses and during exertion.

No known cure exists. Corticosteroids and cytotoxic drugs may be given to suppress inflammation but are usually unsuccessful. Recently, interferon gamma-1b has shown some promise in treating the disease.

Lung transplantation may be successful for younger, otherwise healthy individuals.

## Special Considerations

- ◆ Explain all diagnostic tests to the patient, who may experience anxiety and frustration about the many tests required to establish the diagnosis.
- ◆ Monitor oxygenation at rest and with exertion. The physician may prescribe one oxygen flow rate for use when the patient is at rest and a higher one for use during exertion to maintain adequate oxygenation. Instruct the patient to increase oxygen flow rate to the appropriate level for exercise.
- ◆ As IPF progresses, the patient's oxygen requirements will increase. They may need a nonrebreathing mask to supply high oxygen percentages. Eventually, maintaining adequate oxygenation may become impossible despite maximum oxygen flow.
- ◆ Most patients will need oxygen at home. Make appropriate referrals to discharge planners, respiratory care practitioners, and home equipment vendors to ensure continuity of care.
- ◆ Teach breathing, relaxation, and energy conservation techniques to help the patient manage severe dyspnea.
- ◆ Encourage the patient to be as active as possible. Refer them to a pulmonary rehabilitation program.
- ◆ Monitor the patient for adverse reactions to drug therapy.
- ◆ Teach the patient about prescribed medications, especially adverse effects. Teach the patient and their family members infection prevention techniques.
- ◆ Encourage good nutritional habits. Small, frequent meals with high nutritional value may be necessary if dyspnea interferes with eating.

- ◆ Provide emotional support for the patient and their family as they deal with the patient's increasing disability, dyspnea, and probable death. Consult hospice as appropriate.

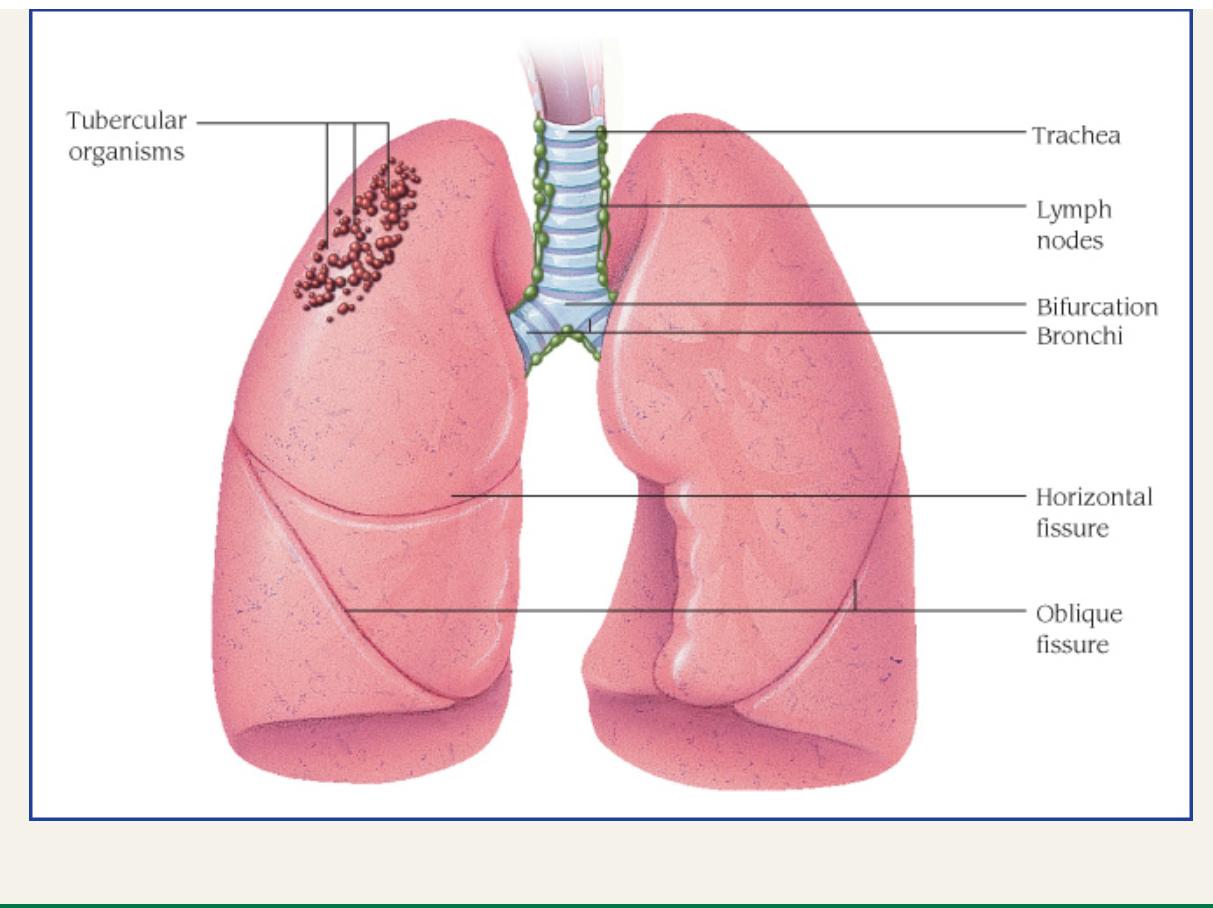
## TUBERCULOSIS

An acute or chronic infection caused by *Mycobacterium tuberculosis*, TB is characterized by pulmonary infiltrates, formation of granulomas with caseation, fibrosis, and cavitation. (See *Understanding tuberculosis invasion*.) People who live in crowded, poorly ventilated conditions and those who are immunocompromised are most likely to become infected. In patients with strains that are sensitive to the usual antitubercular agents, the prognosis is excellent with correct treatment. However, in those with strains that are resistant to two or more of the major antitubercular agents, mortality is 50%.



### PATHOPHYSIOLOGY UNDERSTANDING TUBERCULOSIS INVASION

After infected droplets are inhaled, they enter the lungs and are deposited either in the lower part of the upper lobe or in the upper part of the lower lobe. Leukocytes surround the droplets, which leads to inflammation. As part of the inflammatory response, some mycobacteria are carried off in the lymphatic circulation by the lymph nodes.



## Causes and Incidence

After exposure to *M. tuberculosis*, roughly 5% of infected people develop active TB within 1 year; in the remainder, microorganisms cause a latent infection. The host's immune system usually controls the tubercle bacillus by enclosing it in a tiny nodule (tubercle). The bacillus may lie dormant within the tubercle for years and later reactivate and spread.

Although the primary infection site is the lungs, mycobacteria commonly exist in other parts of the body. Several factors increase the risk of infection reactivation: gastrectomy, uncontrolled diabetes mellitus, Hodgkin lymphoma, leukemia, silicosis, acquired immunodeficiency syndrome, treatment with corticosteroids or immunosuppressants, and advanced age.

Cell-mediated immunity to the mycobacteria, which develops 3 to 6 weeks later, usually contains the infection and arrests the disease. If the infection reactivates, the body's response characteristically leads to caseation—the conversion of necrotic tissue to a cheese-like material. The caseum may localize, undergo fibrosis, or excavate and form cavities, the walls of which are studded with multiplying tubercle bacilli. If this happens, infected caseous

debris may spread throughout the lungs by the tracheobronchial tree. Sites of extrapulmonary TB include the pleurae, meninges, joints, lymph nodes, peritoneum, genitourinary tract, and bowel.

The incidence of TB has been increasing in the United States secondary to homelessness, drug abuse, and human immunodeficiency virus infection. Globally, TB is the leading infectious cause of morbidity and mortality, generating 8 to 10 million new cases each year.

## Pathophysiology

Transmission is by droplet nuclei produced when infected persons cough or sneeze. Persons with a cavitary lesion are particularly infectious because their sputum usually contains 1 to 100 million bacilli per milliliter. If an inhaled tubercle bacillus settles in an alveolus, infection occurs, with alveolocapillary dilation and endothelial cell swelling. Alveolitis results, with replication of tubercle bacilli and influx of polymorphonuclear leukocytes. These organisms spread through the lymph system to the circulatory system and then through the body.

## Complications

- ◆ Respiratory failure
- ◆ Bronchopleural fistulas
- ◆ Pneumothorax
- ◆ Hemorrhage
- ◆ Pleural effusion
- ◆ Pneumonia

## Signs and Symptoms

After an incubation period of 4 to 8 weeks, TB is usually asymptomatic in primary infection but may produce nonspecific symptoms, such as fatigue, weakness, anorexia, weight loss, night sweats, and low-grade fever.

 **ELDER TIP** Fever and night sweats, the typical hallmarks of TB, may not be present in elderly patients, who instead may exhibit a change in activity or weight. Assess older patients carefully.

In reactivation, symptoms may include a cough that produces mucopurulent sputum, occasional hemoptysis, and chest pains.

## Diagnosis

 **CONFIRMING DIAGNOSIS** Diagnostic tests include chest X-rays, a tuberculin skin test, and sputum smears and cultures to identify M. tuberculosis. The diagnosis must be precise because several other diseases (such as lung cancer, lung abscess, pneumoconiosis, and bronchiectasis) may mimic TB.

These procedures aid in diagnosis:

- ◆ Auscultation detects crepitant crackles, bronchial breath sounds, wheezing, and whispered pectoriloquy.
- ◆ Chest percussion detects dullness over the affected area, indicating consolidation or pleural fluid.
- ◆ Chest X-ray shows nodular lesions, patchy infiltrates (mainly in upper lobes), cavity formation, scar tissue, and calcium deposits; however, it may not be able to distinguish active from inactive TB.
- ◆ Tuberculin skin test detects TB infection. Intermediate-strength purified protein derivative or 5 tuberculin units (0.1 mL) are injected intracutaneously on the forearm. The test results are read in 48 to 72 hours; a positive reaction (induration of 5 to 15 mm or more, depending on risk factors) develops 2 to 10 weeks after infection in active and inactive TB. However, severely immunosuppressed patients may never develop a positive reaction.

 **CONFIRMING DIAGNOSIS** Stains and cultures (of sputum, cerebrospinal fluid, urine, drainage from abscess, or pleural fluid) show heat-sensitive, nonmotile, aerobic, acid-fast bacilli.

## Treatment

First-line agents for the treatment of TB are isoniazid (INH), rifampin (RIF), ethambutol (EMB), and pyrazinamide. Latent TB is usually treated with daily INH for 9 months. RIF daily for 4 months may be used for people with latent TB whose contacts are INH resistant. For most adults with active TB, the recommended dosing includes the administration of all four drugs daily for 2 months, followed by 4 months of INH and RIF. Drug therapy must be selected according to patient condition and organism susceptibility. Another first-line drug used for TB is rifapentine. Second-line agents, such as cycloserine, ethionamide, *p*-aminosalicylic acid, streptomycin, and capreomycin, are reserved for special circumstances or drug-resistant strains. Interruption of drug therapy may require initiation of therapy from the beginning of the regimen or additional treatment.

Directly observed therapy (DOT) may be selected or required. In this therapy, an assigned caregiver directly observes the administration of the drug. The goal of DOT is to monitor the treatment regimen and reduce the development of resistant organisms.

## **Special Considerations**

- ◆ Initiate AFB isolation precautions immediately for all patients suspected or confirmed to have TB. AFB isolation precautions include the use of a private room with negative pressure in relation to surrounding areas and a minimum of six air exchanges per hour (air should be exhausted directly to the outside).
- ◆ Continue AFB isolation until there's clinical evidence of reduced infectiousness (substantially decreased cough, fewer organisms on sequential sputum smears).
- ◆ Teach the infectious patient to cough and sneeze into tissues and to dispose of all secretions properly. Place a covered trash can nearby or tape a lined bag to the side of the bed to dispose of used tissues.
- ◆ Instruct the patient to wear a mask when outside his or her room.
- ◆ Visitors and staff members should wear particulate respirators that fit closely around the face when they're in the patient's room.
- ◆ Remind the patient to get plenty of rest. Stress the importance of eating balanced meals to promote recovery. If the patient is anorexic, urge him or her to eat small meals frequently. Record weight weekly.
- ◆ Be alert for adverse effects of medications. Because INH sometimes leads to hepatitis or peripheral neuritis, monitor aspartate aminotransferase and alanine aminotransferase levels. To prevent or treat peripheral neuritis, give pyridoxine (vitamin B<sub>6</sub>), as ordered. If the patient receives EMB, watch for optic neuritis; if it develops, discontinue the drug. If the patient receives RIF, watch for hepatitis and purpura. Observe the patient for other complications, such as hemoptysis.
- ◆ Before discharge, advise the patient to watch for adverse effects from the medication and report them immediately. Emphasize the importance of regular follow-up examinations. Instruct the patient and family concerning the signs and symptoms of recurring TB. Stress the need to follow long-term treatment faithfully.
- ◆ Emphasize to the patient the importance of taking the medications daily as prescribed. The patient may enroll in a supervised administration program

to avoid the development of drug-resistant organisms. (See *Preventing tuberculosis*, page 139.)



## PREVENTION PREVENTING TUBERCULOSIS

The best way to prevent tuberculosis (TB) is early detection to prevent it from becoming active. Hospitalized patients with TB should be isolated from other patients using airborne precautions. Staff members should also use disposable high-efficiency particulate air filter masks, which serve as adequate respiratory protection when caring for patients who are in airborne isolation.

Other ways to prevent the spread of TB include:

- ◆ If a patient has a weakened immune system or has human immunodeficiency virus, it is recommended that they receive annual TB testing. Annual testing is also recommended for healthcare workers, those who work in a prison or a long-term care facility, and those with a substantially increased risk of exposure to the disease.
- ◆ If a patient tests positive for latent TB infection but has no evidence of active TB, he or she may be able to reduce the risk of developing active TB by taking a course of therapy with isoniazid.

To prevent the spread of disease from those with active TB or from those who are receiving treatment, the following recommendations should be followed:

- ◆ Stress the need to maintain the treatment regimen and to not stop or skip doses. When the treatment regimen is stopped, the TB bacteria can mutate and become drug resistant.
- ◆ The patient who is on a treatment regimen is still contagious until he or she has been taking the medications for 2 to 3 weeks. Encouraging the patient to stay indoors and home from school or work is recommended. If the patient must leave home, a mask is recommended during this initial treatment time to lessen the risk of transmission.

## Pneumoconioses

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## SILICOSIS

Silicosis is a progressive disease characterized by nodular lesions that commonly progress to fibrosis. The most common form of pneumoconiosis, silicosis can be classified according to the severity of pulmonary disease and the rapidity of its onset and progression. It usually occurs as a simple asymptomatic illness.

*Acute silicosis* develops after 1 to 3 years in workers exposed to very high concentrations of respirable silica (sand blasters and tunnel workers). *Accelerated silicosis* appears after an average of 10 years of exposure to lower concentrations of free silica. *Chronic silicosis* develops after 20 or more years of exposure to lower concentrations of free silica. (Chronic silicosis is further subdivided into simple and complicated forms.)

The prognosis is good unless the disease progresses into the complicated fibrotic form, which causes respiratory insufficiency and cor pulmonale. It's also associated with pulmonary TB.

### Causes and Incidence

Silicosis results from the inhalation and pulmonary deposition of respirable crystalline silica dust, mostly from quartz. The danger to the worker depends on the concentration of dust in the atmosphere, the percentage of respirable free silica particles in the dust, and the duration of exposure. Respirable particles are less than 10 µm in diameter, but the disease-causing particles deposited in the alveolar space are usually 1 to 3 µm in diameter.

Industrial sources of silica in its pure form include the manufacture of ceramics (flint) and building materials (sandstone). It occurs in mixed form in the production of construction materials (cement). It's found in powder form (silica flour) in paints, porcelain, scouring soaps, and wood fillers as well as in the mining of gold, coal, lead, zinc, and iron. Foundry workers, boiler scalers, and stonemasons are all exposed to silica dust and, therefore, are at high risk for developing silicosis.

The incidence of silicosis has decreased since the Occupational Safety and Health Administration instituted regulations requiring the use of protective equipment that limits the amount of silica dust inhaled.

### Pathophysiology

Nodules result when alveolar macrophages ingest silica particles, which they're unable to process. As a result, the macrophages die and release

proteolytic enzymes into the surrounding tissue. The subsequent inflammation attracts other macrophages and fibroblasts into the region to produce fibrous tissue and wall off the reaction. The resulting nodule has an onionskin appearance when viewed under a microscope. Nodules develop adjacent to terminal and respiratory bronchioles, concentrate in the upper lobes, and are commonly accompanied by bullous changes in both lobes. If the disease process doesn't progress, minimal physiologic disturbances and no disability occur. Occasionally, however, the fibrotic response accelerates, engulfing and destroying large areas of the lung (progressive massive fibrosis or conglomerate lesions). Fibrosis may continue even after exposure to dust has ended.

## Complications

- ◆ Pulmonary fibrosis
- ◆ Cor pulmonale
- ◆ Ventricular or respiratory failure
- ◆ Pulmonary TB

## Signs and Symptoms

Initially, silicosis may be asymptomatic or may produce dyspnea on exertion, usually attributed to being “out of shape” or “slowing down.” If the disease progresses to the chronic and complicated stage, dyspnea on exertion worsens, and other symptoms—usually tachypnea and an insidious dry cough that's most pronounced in the morning—appear.

Progression to the advanced stage causes dyspnea on minimal exertion, worsening cough, and pulmonary hypertension, which in turn leads to right-sided heart failure and cor pulmonale. Patients with silicosis have a high incidence of active TB, which should be considered when evaluating patients with this disease. CNS changes—confusion, lethargy, and a decrease in the rate and depth of respiration as the  $\text{Paco}_2$  increases—also occur in advanced silicosis.

Other clinical features include malaise, disturbed sleep, and hoarseness. The severity of these symptoms may not correlate with chest X-ray findings or the results of PFTs.

## Diagnosis

The patient history reveals occupational exposure to silica dust. The physical examination is normal in simple silicosis; in chronic silicosis with conglomerate lesions, it may reveal decreased chest expansion, diminished intensity of breath sounds, areas of hyporesonance and hyperresonance, fine to medium crackles, and tachypnea.

In simple silicosis, chest X-rays show small, discrete, nodular lesions distributed throughout both lung fields but typically concentrated in the upper lung zones; the hilar lung nodes may be enlarged and exhibit “eggshell” calcification. In complicated silicosis, X-rays show one or more conglomerate masses of dense tissue.

PFTs show:

- ◆ FVC—reduced in complicated silicosis
- ◆ FEV<sub>1</sub>—reduced in obstructive disease (emphysematous areas of silicosis); reduced in complicated silicosis, but ratio of FEV<sub>1</sub> to FVC is normal or high
- ◆ Maximal voluntary ventilation—reduced in restrictive and obstructive diseases
- ◆ CO<sub>2</sub> diffusing capacity—reduced when fibrosis destroys alveolar walls and obliterates pulmonary capillaries or when fibrosis thickens the alveolocapillary membrane

## Treatment

The goal of treatment is to relieve respiratory symptoms, to manage hypoxemia and cor pulmonale, and to prevent respiratory tract irritation and infections. Treatment also includes careful observation for the development of TB. Respiratory symptoms may be relieved through daily use of inhaled bronchodilators and increased fluid intake (at least 3 qt [3 L] daily). Steam inhalation and chest physiotherapy techniques, such as controlled coughing and segmental bronchial drainage with chest percussion and vibration, help clear secretions. In severe cases, it may be necessary to administer oxygen by cannula or mask (1 to 2 L/minute) for the patient with chronic hypoxemia or by mechanical ventilation if arterial oxygen can't be maintained above 40 mm Hg. Respiratory infections require prompt administration of antibiotics.

## Special Considerations

- ◆ Teach the patient to prevent infections by avoiding crowds and persons with respiratory infections and by receiving influenza and pneumococcal

vaccines.

- ◆ Increase exercise tolerance by encouraging regular activity. Advise the patient to plan his or her daily activities to decrease the work of breathing. The patient should be instructed to pace himself or herself, rest often, and generally move slowly through his or her daily routine.

## ASBESTOSIS

Asbestosis is a form of pneumoconiosis characterized by diffuse interstitial fibrosis. It can develop as long as 15 to 20 years after regular exposure to asbestos has ended. Asbestos also causes pleural plaques and mesotheliomas of pleura and the peritoneum. A potent co-carcinogen, asbestos increases the risk of lung cancer in cigarette smokers.

### Causes and Incidence

Asbestosis results from the inhalation of respirable asbestos fibers (50 µm or more in length and 0.5 µm or less in diameter), which assume a longitudinal orientation in the airway and move in the direction of airflow. The fibers penetrate respiratory bronchioles and alveolar walls. Sources include the mining and milling of asbestos, the construction industry, and the fireproofing and textile industries. Asbestos was also used in the production of paints, plastics, and brake and clutch linings.

Asbestos-related diseases develop in families of asbestos workers as a result of exposure to fibrous dust shaken off workers' clothing at home. Such diseases develop in the general public as a result of exposure to fibrous dust or waste piles from nearby asbestos plants, but exposures for occupants of typical buildings are quite low and not in a range associated with asbestosis.

Inhaled fibers become encased in a brown, protein-like sheath rich in iron (ferruginous bodies or asbestos bodies), found in sputum and lung tissue. Interstitial fibrosis develops in lower lung zones, causing obliterative changes in lung parenchyma and pleurae. Raised hyaline plaques may form in parietal pleura, diaphragm, and pleura contiguous with the pericardium.

Asbestosis occurs in 4 of every 10,000 people.

### Pathophysiology

Asbestos fibers penetrate the pleura tissues and are phagocytosed permanently within the lungs. This sets up cycles of cellular events and the release of cytokines. The initial irritation occurs in the alveoli and initiates a surge of asbestos-activated macrophages. These specified macrophages then

produce multiple varieties of growth factors that interact to produce fibroblast proliferation.

## Complications

- ◆ Pulmonary fibrosis
- ◆ Respiratory failure
- ◆ Pulmonary hypertension
- ◆ Cor pulmonale

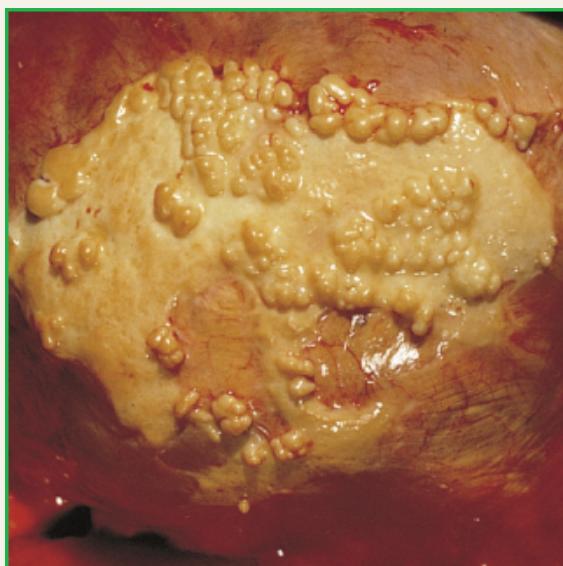
## Signs and Symptoms

Clinical features may appear before chest X-ray changes. The first symptom is usually dyspnea on exertion, typically after 10 years' exposure. As fibrosis extends, dyspnea on exertion increases until, eventually, dyspnea occurs even at rest. Advanced disease also causes a dry cough (may be productive in smokers), chest pain (commonly pleuritic), recurrent respiratory infections, and tachypnea. (See *A close look at asbestosis*.)

## A Close Look at Asbestosis

After years of exposure to asbestos, healthy lung tissue progresses from simple asbestosis to massive pulmonary fibrosis, as shown below.

**Simple asbestosis**



**Progressive massive pulmonary fibrosis**



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Cardiovascular complications include pulmonary hypertension, right ventricular hypertrophy, and cor pulmonale. Finger clubbing commonly occurs.

## Diagnosis

The patient history reveals occupational, family, or neighborhood exposure to asbestos fibers. Physical examination reveals characteristic dry crackles at lung bases. Chest X-rays show fine, irregular, and linear diffuse infiltrates; extensive fibrosis results in a “honeycomb” or “ground-glass” appearance. X-rays may also show pleural thickening and calcification, with bilateral obliteration of costophrenic angles. In later stages, an enlarged heart with a classic “shaggy” heart border may be evident. CT scan of the lungs also aids in diagnosis.

PFTs show:

- ◆ Vital capacity, FVC, and TLC—decreased
- ◆ FEV<sub>1</sub>—decreased or normal
- ◆ Carbon monoxide diffusing capacity—reduced when fibrosis destroys alveolar walls and thickens alveolocapillary membranes

ABG analysis reveals:

- ◆  $\text{PaO}_2$ —decreased
- ◆  $\text{PaCO}_2$ —low due to hyperventilation

## Treatment

The goal of treatment is to relieve respiratory symptoms and, in advanced disease, manage hypoxemia and cor pulmonale. Respiratory symptoms may be relieved by chest physiotherapy techniques, such as controlled coughing and segmental bronchial drainage, chest percussion, and vibration. Aerosol therapy, inhaled mucolytics, and increased fluid intake (at least 3 qt [3 L] daily) may also relieve symptoms.

Diuretics, cardiac glycosides, and salt restriction may be indicated for patients with cor pulmonale. Hypoxemia requires oxygen administration by cannula or mask (1 to 2 L/minute) or by mechanical ventilation if arterial oxygen can't be maintained above 40 mm Hg. Respiratory infections require prompt administration of antibiotics.

## Special Considerations

- ◆ Teach the patient to prevent infections by avoiding crowds and persons with infections and by receiving influenza and pneumococcal vaccines.
- ◆ Improve the patient's ventilatory efficiency by encouraging physical reconditioning, energy conservation in daily activities, and relaxation techniques.

## COAL WORKER'S PNEUMOCONIOSIS

A progressive nodular pulmonary disease, coal worker's pneumoconiosis (CWP) occurs in two forms. Simple CWP is characterized by small lung opacities; in complicated CWP, also known as *progressive massive fibrosis*, masses of fibrous tissue occasionally develop in the patient's lungs. The risk of developing CWP (also known as *black lung disease*, *coal miner's disease*, *miner's asthma*, *anthracosis*, and *anthracosilicosis*) depends on the duration of exposure to coal dust (usually 15 years or longer), intensity of exposure (dust count and particle size), location of the mine, silica content of the coal (anthracite coal has the highest silica content), and the worker's susceptibility.

The prognosis varies. Simple asymptomatic disease is self-limiting, although progression to complicated CWP is more likely if CWP begins after a relatively short period of exposure. Complicated CWP may be disabling, resulting in severe ventilatory failure and cor pulmonale.

## **Causes and Incidence**

CWP is caused by the inhalation and prolonged retention of respirable coal dust particles (less than 5  $\mu\text{m}$  in diameter). Simple CWP results in the formation of macules (accumulations of macrophages laden with coal dust) around the terminal and respiratory bronchioles, surrounded by a halo of dilated alveoli. Macule formation leads to atrophy of supporting tissue, causing permanent dilation of small airways (focal emphysema).

Simple disease may progress to complicated CWP, involving one or both lungs. In this form of the disease, fibrous tissue masses enlarge and coalesce, causing gross distortion of pulmonary structures (destruction of vasculature alveoli and airways).

The incidence of CWP is highest among anthracite coal miners in the eastern United States.

## **Pathophysiology**

When coal dust particles are inhaled into the lung bronchioles, the carbon is phagocytosed and transported by macrophages and microciliary into the mucus. An immune response is triggered as the coal-filled macrophages accumulate in the alveoli. Reticulin is secreted from the fibroblasts which then entrap the macrophages and become strangulated from the resultant interstitial fibrosis.

## **Complications**

- ◆ Pulmonary hypertension
- ◆ Pulmonary TB
- ◆ Cor pulmonale

## **Signs and Symptoms**

Simple CWP produces no symptoms, especially in nonsmokers. Symptoms of complicated CWP include exertional dyspnea and a cough that occasionally produces inky-black sputum (when fibrotic changes undergo avascular necrosis and their centers cavitate). Other clinical features of CWP include increasing dyspnea and a cough that produces milky, gray, clear, or coal-flecked sputum. Recurrent bronchial and pulmonary infections produce yellow, green, or thick sputum.

Complications include pulmonary hypertension, right ventricular hypertrophy, cor pulmonale, and pulmonary TB. In cigarette smokers, chronic

bronchitis and emphysema may also complicate the disease.

## Diagnosis

The patient history reveals exposure to coal dust. Physical examination shows barrel chest, hyperresonant lungs with areas of dullness, diminished breath sounds, crackles, rhonchi, and wheezes. In *simple CWP*, chest X-rays show small opacities (less than 10 mm in diameter). These may be present in all lung zones but are more prominent in the upper lung zones. In *complicated CWP*, one or more large opacities (1 to 5 cm in diameter), possibly exhibiting cavitation, are seen.

PFTs show:

- ◆ Vital capacity—normal in simple CWP but decreased with complicated CWP
- ◆ FEV<sub>1</sub>—decreased in complicated disease
- ◆ Residual volume and TLC—normal in simple CWP; decreased in complicated CWP
- ◆ Carbon monoxide diffusing capacity—significantly decreased in complicated CWP as alveolar septae are destroyed and pulmonary capillaries obliterated
- ◆ Paco<sub>2</sub>—may be increased with concomitant COPD

## Treatment

There's no specific treatment. The goal of treatment is to relieve respiratory symptoms, manage hypoxia and cor pulmonale, and avoid respiratory tract irritants and infections. Treatment also includes careful observation for the development of TB. Chest physiotherapy techniques, such as controlled coughing and segmental bronchial drainage combined with chest percussion and vibration, help remove secretions.

Other measures include increased fluid intake (at least 3 qt [3 L] daily) and respiratory therapy techniques, such as aerosol therapy, inhaled mucolytics, and intermittent positive-pressure breathing. Diuretics, cardiac glycosides, and salt restriction may be indicated in cor pulmonale. In severe cases, it may be necessary to administer oxygen for hypoxemia by cannula or mask (1 to 2 L/minute) if the patient has chronic hypoxia; mechanical ventilation is utilized if Pao<sub>2</sub> can't be maintained above 40 mm Hg. Respiratory infections require prompt administration of antibiotics.

## Special Considerations

- ◆ Teach the patient to prevent infections by avoiding crowds and persons with respiratory infections and by receiving pneumococcal vaccine polyvalent and annual influenza vaccines.
- ◆ Encourage the patient to stay active to avoid a deterioration in his or her physical condition but to pace activities and practice relaxation techniques.

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# 10

## Metabolic and Nutritional Disorders

### Introduction

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Metabolism is the physiologic process that allows cells to transform food into energy and continually rebuild body cells. Metabolism has two phases: catabolism and anabolism. In *catabolism*, the energy-producing phase of metabolism, the body breaks down large food molecules into smaller ones; in *anabolism*, the tissue-building phase, the body converts small molecules into larger ones (such as antibodies to keep the body capable of fighting infection). Both phases are accomplished by means of a chemical process using energy. A wide range of nutrients is metabolized to meet the body's needs. (See *Essential nutrients and their functions*, page 487.)



**ELDER TIP** *A person's protein, vitamin, and mineral requirements usually remain the same as they age, although calorie needs decline. Diminished activity may lower energy requirements by almost 200 calories/day for men and women ages 51 to 75, 400 calories/day for women older than age 75, and 500 calories/day for men older than age 75.*

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### Essential nutrients and their functions

Nutrients are required for the body to work properly and avoid disease.

| <b>Nutrients</b>               | <b>Functions</b>  |
|--------------------------------|---|
| Carbohydrates                  | ◆ Energy source   |
| Fats and essential fatty acids | ◆ Energy source; essential for growth, normal skin, and membranes   |
| Proteins and amino acids       | ◆ Synthesis of all body proteins, growth, and tissue maintenance  |
| <b>Water-soluble vitamins</b>  |   |
| ◆ Ascorbic acid (C)            | ◆ Collagen synthesis, wound healing, antioxidation<br>◆ Coenzyme in carbohydrate metabolism   |
| ◆ Thiamine (B1)                | ◆ Coenzyme in energy metabolism   |
| ◆ Riboflavin (B2)              | ◆ Coenzyme in carbohydrate, fat, energy metabolism, and tissue metabolism   |
| ◆ Niacin                       | ◆ Deoxyribonucleic acid (DNA) and ribonucleic acid synthesis; erythrocyte formation   |
| ◆ Vitamin B12                  | ◆ Coenzyme in amino acid metabolism; heme and hemoglobin formation; DNA synthesis; lowering homocysteine levels   |
| ◆ Folic acid                   |   |
| <b>Fat-soluble vitamins</b>    |   |
| ◆ Vitamin A                    | ◆ Vision in dim light, mucosal epithelium integrity, tooth development, endocrine function  |
| ◆ Vitamin D                    |   |
| ◆ Vitamin E                    | ◆ Regulation of calcium and phosphate absorption and metabolism; renal phosphate clearance; musculoskeletal function  |
| ◆ Vitamin K                    | ◆ Antioxidation; essential for muscle, liver, and red blood cell integrity<br>◆ Blood clotting (catalyzes synthesis of prothrombin by liver); skeletal function |

## CARBOHYDRATES: PRIMARY ENERGY SOURCE

The body gets most of its energy by metabolizing carbohydrates, especially glucose. Glucose catabolism proceeds in three phases:

- ◆ *Glycolysis*, a series of chemical reactions, converts glucose molecules into pyruvic or lactic acid.
- ◆ The *citric acid cycle* removes ionized hydrogen atoms from pyruvic acid and produces carbon dioxide.
- ◆ *Oxidative phosphorylation* traps energy from the hydrogen electrons and combines the hydrogen ions and electrons with oxygen to form water and the common form of biological energy, adenosine triphosphate (ATP).

Other essential processes in carbohydrate metabolism include glycogenesis—the formation of glycogen, a storage form of glucose—which occurs when cells become saturated with glucose-6-phosphate (an intermediate product of glycolysis); glycogenolysis, the reverse process,

which converts glycogen into glucose-6-phosphate in muscle cells and liberates free glucose in the liver; and gluconeogenesis, or “new” glucose formation from protein amino acids or fat glycerols.

A complex interplay of hormonal and neural controls regulates glucose metabolism. Hormone secretions of five endocrine glands dominate this regulatory function:

- ◆ Alpha cells of the islets of Langerhans secrete glucagon, which increases the blood glucose level by stimulating phosphorylase activity to accelerate liver glycogenolysis.
- ◆ Beta cells of the islets of Langerhans secrete the glucose-regulating hormone insulin, which assists in glucose transport across cell membranes and storage of excess glucose as fat.
- ◆ The adrenal medulla, as a physiologic response to stress, secretes epinephrine, which stimulates liver and muscle glycogenolysis to increase the blood glucose level.
- ◆ Corticotropin and glucocorticoids also increase blood glucose levels. Glucocorticoids accelerate gluconeogenesis by promoting the flow of amino acids to the liver, where they’re synthesized into glucose.
- ◆ Human growth hormone (hGH) limits the fat storage and favors fat catabolism; consequently, it inhibits carbohydrate catabolism and thus raises blood glucose levels.
- ◆ Thyroid-stimulating hormone and thyroid hormone have mixed effects on carbohydrate metabolism and may raise or lower blood glucose levels.

## FATS: CATABOLISM AND ANABOLISM

The breaking up of triglycerides—*lipolysis*—yields fatty acids and glycerol. Beta oxidation breaks down fatty acids into acetyl coenzyme A, which can then enter the citric acid cycle; glycerol can also undergo gluconeogenesis or enter the glycolytic pathways to produce energy. Conversely, *lipogenesis* is the chemical formation of fat from excess carbohydrates and proteins or from the fatty acids and glycerol products of lipolysis. Adipose tissue is the primary storage site for excess fat and thus is the greatest source of energy reserve. Certain unsaturated fatty acids are necessary for the synthesis of vital body compounds. Because the body can't produce these essential fatty acids, they must be provided through diet. Insulin, hGH, catecholamines,

corticotropin, and glucocorticoids control fat metabolism in an inverse relationship with carbohydrate metabolism; large amounts of carbohydrates promote fat storage, and deficiency of available carbohydrates promotes fat breakdown for energy needs.

## PROTEINS: ANABOLISM

The primary process in protein metabolism is anabolism. Catabolism is relegated to a supporting role in protein metabolism—a reversal of the roles played by these two processes in carbohydrate and fat metabolisms. By synthesizing proteins—the tissue-building foods—the body derives substances essential for life (such as plasma proteins) and can reproduce, control cell growth, and repair itself. However, when carbohydrates or fats are unavailable as energy sources, or when energy demands are exceedingly high, protein catabolism converts protein into an available energy source. Protein metabolism consists of many processes, including:

- ◆ *Deamination*—a catabolic and energy-producing process occurring in the liver with the splitting off of the amino acid to form ammonia and a keto acid
- ◆ *Transamination*—anabolic conversion of keto acids to amino acids
- ◆ *Urea formation*—a catabolic process occurring in the liver, producing urea, the end product of protein catabolism

The male hormone testosterone and hGH stimulate protein anabolism; corticotropin prompts secretion of glucocorticoids, which, in turn, facilitate protein catabolism. Normally, the rate of protein anabolism equals the rate of protein catabolism—a condition known as *nitrogen balance* (because ingested nitrogen equals nitrogen waste excreted in urine, feces, and sweat). When excessive catabolism causes the amount of nitrogen excreted to exceed the amount ingested, a state of *negative nitrogen balance* exists—usually the result of starvation and cachexia or surgical stress.

## FLUID AND ELECTROLYTE BALANCE

A critical component of metabolism is fluid and electrolyte balance. Water is an essential body substance and constitutes almost 60% of an adult's body weight and more than 75% of a neonate's body weight. In both older and obese adults, the ratio of water to body weight drops; children and lean people have a higher proportion of water in their bodies.

Body fluids can be classified as intracellular (or cellular) or extracellular. Intracellular fluid constitutes about 40% of total body weight and 60% of all body fluid; it contains large quantities of potassium and phosphates but very little sodium and chloride. Conversely, extracellular fluid (ECF) contains mostly sodium and chloride but very little potassium and phosphates. Incorporating interstitial, cerebrospinal, intraocular, and gastrointestinal (GI) fluids and plasma, ECF supplies cells with nutrients and other substances needed for cellular function. The many components of body fluids have the important function of preserving osmotic pressure and acid–base and anion–cation balance.

Homeostasis is a stable state—the equilibrium of chemical and physical properties of body fluid. Body fluids contain two kinds of dissolved substances: those that dissociate in solution (electrolytes) and those that don't. For example, glucose, when dissolved in water, doesn't break down into smaller particles; but sodium chloride dissociates in solution into sodium cations (+) and chloride anions (−). The composition of these electrolytes in body fluids is electrically balanced so the positively charged ions (cations: sodium, potassium, calcium, and magnesium) equal the negatively charged ions (anions: chloride, bicarbonate, sulfate, phosphate, proteinate, and carbonic and other organic acids). Although these particles are present in relatively low concentrations, any deviation from their normal levels can have profound physiologic effects.



**ELDER TIP** Institutionalized older people are at particularly high risk for dehydration because of their diminished thirst perception and any combination of physical, cognitive, speech, mobility, and visual impairment.

In homeostasis—an ever-changing but balanced state—water and electrolytes and other solutes move continually between cellular and extracellular compartments. Such motion is made possible by semipermeable membranes that allow diffusion, filtration, and active transport. *Diffusion* refers to the movement of particles or molecules from an area of greater concentration to one of lesser concentration. Normally, particles move randomly and constantly until the concentrations within given solutions are equal. Diffusion also depends on permeability, electrical gradient, and pressure gradient. Particles, however, can't diffuse against any of these gradients without energy and a carrier substance (active transport).

ATP is released from cells to aid particles needing energy to pass through the cell membrane.

The diffusion of water from a solution of low concentration to one of high concentration is called *osmosis*. The pressure that develops when a selectively permeable cell membrane separates solutions of different strengths of concentrations is known as *osmotic pressure*, expressed in terms of osmoles or milliosmoles (mOsm). Osmotic activity is described in terms of *osmolality*—the osmotic pull exerted by all particles per unit of water, expressed in mOsm/kg of water—or *osmolarity*, when expressed in mOsm/L of solution.

The normal range of body fluid osmolality is 285 to 295 mOsm/kg. Solutions of 50 mOsm above or below the high and low points of this normal range exert little or no osmotic effect (iso-osmolality). A solution below 240 mOsm contains a lower particle concentration than plasma (hypo-osmolar), whereas a solution over 340 mOsm has a higher particle concentration than plasma (hyperosmolar).

Rapid I.V. administration of iso-osmolar solutions to patients who are debilitated, are very old or very young, or have cardiac or renal insufficiency could lead to ECF volume overload and induce pulmonary edema and heart failure because particulate concentration is the same as plasma, so fluid shifting into and out of cells will occur.

Continuous I.V. administration of hypo-osmolar solutions decreases serum osmolality and leads to excess intracellular fluid volume (water intoxication), whereas continuous I.V. administration of hyperosmolar solutions results in intracellular dehydration, increased serum osmolality and, eventually, ECF volume deficit due to excessive urinary excretion. These states occur because of fluid diffusion and the cell's attempt to balance the particulate concentrations inside and outside the cell.

## **REGULATION OF PH**

Primarily through the complex chemical regulation of carbonic acid by the lungs and of base bicarbonate by the kidneys, the body maintains the hydrogen ion concentration to keep the ECF pH between 7.35 and 7.45. Nutritional deficiency or excess, disease, injury, or metabolic disturbance can interfere with normal homeostatic mechanisms and raise pH (acidosis) or lower it (alkalosis).

## ASSESSING HOMEOSTASIS

The goal of metabolism and homeostasis is to maintain the complex environment of ECF—the plasma—which nourishes and supports every body cell. This special environment is subject to multiple interlocking influences and readily reflects any disturbance in nutrition, chemical or fluid content, and osmotic pressure. Such disturbances can be detected by various laboratory tests. For example, measurements of albumin, prealbumin, and other blood proteins; electrolyte concentration; enzyme and antibody levels; and urine and blood chemistry levels (lipoproteins, glucose, blood urea nitrogen [BUN], creatinine, and creatinine-height index [CHI]) reflect the state of metabolism, homeostasis, and nutrition throughout the body. (See *Laboratory tests: Assessing nutritional status*, page 490.) Results of such laboratory tests, of course, supplement the information obtained from dietary history and physical examination—which offer gross clinical information about the quality, quantity, and efficiency of metabolic processes. To support clinical information, anthropometry, height-weight ratio, and skinfold thickness determinations specifically define tissue nutritional status.

### Laboratory tests: Assessing nutritional status

Blood and urine tests provide the most precise data about nutritional status, often revealing nutritional problems before they're clinically apparent. The list below explains some common tests and what their results mean.

#### Serum Vitamins and Minerals

Vitamin and mineral deficiencies commonly screened for include deficiencies in A, B, B<sub>12</sub>, folic acid, ascorbic acid, beta-carotene, riboflavin and, sometimes, zinc, calcium, magnesium, iron, and other minerals.

#### Serum Nutrients

Glucose levels help assess suspected diabetes or hypoglycemia. Glucose may be elevated with stress, acromegaly, Cushing syndrome, corticosteroid use, liver disease, sepsis, or overfeeding. Glucose may be

decreased with fluid overload, adrenal insufficiency, liver disease, severe sepsis, insulinoma, or pancreatic disorders. Cholesterol and triglyceride levels help differentiate the type of hyperlipoproteinemia.

### Nitrogen Balance

A negative nitrogen balance indicates inadequate intake of protein or calories.

### Hemoglobin and Hematocrit

Decreased levels can occur in protein-calorie malnutrition, iron deficiency, overhydration, hemorrhage, and hemolytic disease; elevated levels in dehydration, polycythemia, and folate and vitamin B<sub>12</sub> deficiency.

### Serum Albumin

Reduced levels may indicate overhydration or visceral protein depletion because of GI disease, liver disease, or nephrotic syndrome. Elevated levels occur in dehydration.

### Creatinine-Height Index (Chi)

This calculated value reflects muscle mass and estimates muscle protein depletion. Reduced CHI may indicate protein-calorie malnutrition or impaired renal function.

### Serum Prealbumin

This carrier protein for thyroxine is a sensitive indicator of visceral protein.

### Total Lymphocyte Count

This provides an indication of immune status. Counts are low in malnutrition and acquired immunodeficiency syndrome.

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The following measures can help you maintain your patient's homeostasis:

- ◆ Obtain a complete dietary history and nutritional assessment, including weight history and GI symptoms, to determine if carbohydrate, fat,

protein, vitamin, mineral, and water intake are adequate for energy production and for tissue repair and growth. Remember that during periods of rapid tissue synthesis (growth, pregnancy, healing), protein needs increase.

- ◆ Consult a dietitian about any patient who may be malnourished because of malabsorption syndromes, renal or hepatic disease, or clear-liquid diets or who may possibly receive nothing by mouth for more than 5 days. Planned meals that provide adequate carbohydrates, fats, and protein are necessary for convalescence. Supplementary carbohydrates are often needed to spare protein and achieve a positive nitrogen balance.
- ◆ Accurately record intake and output to assess fluid balance (this includes intake of oral liquids or I.V. solutions and urine, gastric, and stool output).
- ◆ Weigh the patient daily—at the same time, with the same-type clothing, and on the same scale. Remember, a weight loss of 2.2 lb (1 kg) is equivalent to the loss of 1 L of fluid.
- ◆ Observe the patient closely for insensible water or unmeasured fluid losses (such as through diaphoresis). Remember, fluid loss from the skin and lungs (normally 900 mL/day) can reach as high as 2,000 mL/day from hyperventilation or tachypnea, thus increasing insensible water losses.



**ELDER TIP** Teach elderly patients and others vulnerable to fluid imbalances the importance of maintaining adequate fluid intake.

- ◆ Recognize I.V. solutions that are hypo-osmolar, such as 0.45% NaCl (half-normal saline solution). Iso-osmolar solutions include normal saline solution (0.9% NaCl), 5% dextrose in 0.2% NaCl, Ringer solutions, and 5% dextrose in water. (The latter acts like a hypotonic solution because dextrose is quickly metabolized, leaving only free water.) Hyperosmolar solutions include 5% dextrose in normal saline solution, 10% dextrose in water, and 5% dextrose in Ringer lactate solution.
- ◆ When continuously administering hypo-osmolar solutions, watch for signs of water intoxication: headaches, behavior changes (confusion or disorientation), nausea, vomiting, rising blood pressure, and falling pulse rate.

- When continuously administering hyperosmolar solutions, be alert for signs of hypovolemia: thirst, dry mucous membranes, slightly falling blood pressure, rising pulse rate and respirations, low-grade fever ( $99^{\circ}\text{ F}$  [ $37.2^{\circ}\text{ C}$ ]), and elevated hematocrit, hemoglobin, and BUN levels.
- Administer fluid cautiously, especially to the patient with cardiopulmonary or renal disease, and watch for signs of overhydration: constant and irritating cough, dyspnea, moist crackles, rising central venous pressure, and pitting edema (late sign). When the patient is in an upright position, neck and hand vein engorgement is a sign of fluid overload.



**ELDER TIP** Many older patients take drugs to treat a variety of conditions. Remember that drugs can affect the patient's nutritional status by altering nutrient absorption, metabolism, utilization, or excretion. Likewise, various foods, beverages, and mineral or vitamin supplements can affect the absorption and effectiveness of drugs. Be aware of these potential interactions when evaluating the patient's medication regimen and nutritional status.

## Nutritional Imbalance

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### VITAMIN A DEFICIENCY

A fat-soluble vitamin absorbed in the GI tract, vitamin A maintains epithelial tissue and retinal function. Consequently, deficiency of this vitamin may result in night blindness, decreased color adjustment, keratinization of epithelial tissue, and poor bone growth. Healthy adults have adequate vitamin A reserves to last up to a year; children often don't.

#### Causes and Incidence

Vitamin A deficiency usually results from inadequate intake of foods high in vitamin A (liver, kidney, butter, milk, cream, cheese, and fortified margarine) or carotene, a precursor of vitamin A found in dark green leafy vegetables and yellow or orange fruits and vegetables. (Six milligrams of beta-carotene is equal to 1 mg of vitamin A.) The recommended daily allowance for vitamin A is 3,000 IU for adult males and 2,310 IU for adult females.

Less common causes of vitamin A deficiency include:

- ◆ malabsorption due to celiac disease, sprue, cirrhosis, obstructive jaundice, cystic fibrosis, giardiasis, or habitual use of mineral oil as a laxative
- ◆ massive urinary excretion caused by cancer, tuberculosis, pneumonia, nephritis, or urinary tract infection
- ◆ decreased storage and transport of vitamin A due to hepatic disease

Each year, more than 80,000 people worldwide—mostly children in underdeveloped countries—lose their sight from severe vitamin A deficiency. This condition is rare in the United States, although many disadvantaged children have substandard levels of vitamin A. With therapy, the chance of reversing symptoms of night blindness and milder conjunctival changes is excellent. When corneal damage is present, emergency treatment is necessary.

## Pathophysiology

Once ingested, provitamins A are released from proteins in the stomach. These retinyl esters are then hydrolyzed to retinol in the small intestine, because retinol is more efficiently absorbed. Carotenoids are cleaved in the intestinal mucosa into molecules of retinaldehyde, which is subsequently reduced to retinol and then esterified to retinyl esters. The retinyl esters of retinoid and carotenoid origin are transported via micelles in the lymphatic drainage of the intestine to the blood and then to the liver as components of chylomicrons. In the body, 50% to 80% of vitamin A is stored in the liver, where it is bound to the cellular retinol-binding protein (RBP). The remaining vitamin A is deposited into adipose tissue, the lungs, and the kidneys as retinyl esters, most commonly as retinyl palmitate.

Vitamin A can be mobilized from the liver to peripheral tissue by a process of deesterification of the retinyl esters. In blood, vitamin A is bound to RBP, which transports it as a complex with transthyretin. The hepatic synthesis of RBP is dependent on the presence of zinc and amino acids to maintain its narrow serum range of 40 to 50 µg/dL. Through a receptor-mediated process, the retinol is taken up by the peripheral tissues from the RBP-transthyretin complex.

## Complication

- ◆ Corneal damage

## Signs and Symptoms

Typically, the first symptom of vitamin A deficiency is night blindness (nyctalopia), which usually becomes apparent when the patient enters a dark place or is caught in the glare of oncoming headlights while driving at night. This condition can progress to xerophthalmia, or drying of the conjunctivas, with development of gray plaques (Bitot spots); if unchecked, perforation, scarring, and blindness may result. Keratinization of epithelial tissue causes dry, scaly skin; follicular hyperkeratosis; and shrinking and hardening of the mucous membranes, possibly leading to infections of the eyes and the respiratory or genitourinary tract. An infant with severe vitamin A deficiency shows signs of failure to thrive and apathy, along with dry skin and corneal changes, which can lead to ulceration and rapid destruction of the cornea.

## Diagnosis

Dietary history and typical ocular lesions suggest vitamin A deficiency. Carotene levels less than 40 µg/dL also suggest vitamin A deficiency, but they vary with seasonal ingestion of fruits and vegetables.



**CONFIRMING DIAGNOSIS** *A serum level of vitamin A that falls below 10 µg/dL confirms the diagnosis. Levels between 10 and 19 µg/dL are also considered low, but the patient isn't likely to have developed significant symptoms.*

## Treatment

Mild conjunctival changes or night blindness requires vitamin A replacement in the form of cod liver oil or halibut liver oil. Acute deficiency requires aqueous vitamin A solution I.M., or oral tablets, especially when corneal changes have occurred. Therapy for underlying biliary obstruction consists of administration of bile salts; for pancreatic insufficiency, pancreatin, or pancrelipase. Dry skin responds well to cream-based or petroleum-based products.

In patients with chronic malabsorption of fat-soluble vitamins, and in those with low dietary intake, prevention of vitamin A deficiency requires aqueous I.V. supplements or an oral water-miscible preparation.

## Special Considerations

- Administer oral vitamin A supplements with or after meals or parenterally, as indicated. Watch for signs of hypercarotenemia (orange coloration of the skin and eyes) and hypervitaminosis A (rash, hair loss, anorexia, transient hydrocephalus, and vomiting in children; bone pain, hepatosplenomegaly, diplopia, and irritability in adults). If these signs occur, discontinue supplements and notify the physician immediately. (Hypercarotenemia is relatively harmless; hypervitaminosis A may be toxic.)



**PREVENTION** Because vitamin A deficiency usually results from dietary insufficiency, provide nutritional counseling. Tell the patient that vitamin A comes from animal sources, such as eggs, meat, milk, cheese, cream, liver, kidney, and cod and halibut fish oil, but that healthier choices, such as carrots, pumpkins, sweet potatoes, and most dark green, leafy vegetables are good sources of beta-carotene, vitamin A's precursor form. Instruct the patient to choose intense-colored fruit or vegetables, which have high beta-carotene content. Provide referrals to appropriate community agencies if necessary.

## VITAMIN B DEFICIENCIES

Vitamin B complex is a group of water-soluble vitamins essential to normal metabolism, cell growth, and blood formation. (See *Recommended daily allowance of B complex vitamins*.) The most common deficiencies involve thiamine (B<sub>1</sub>), riboflavin (B<sub>2</sub>), niacin (B<sub>3</sub>), pyridoxine (B<sub>6</sub>), and cobalamin (B<sub>12</sub>).

### Recommended daily allowance of B complex vitamins

| <b>Vitamin 70)</b> | <b>Men (age 23 to 70)</b> | <b>Women (age 23 to 70)</b> | <b>Infants</b> | <b>Children (age 1 to 18)</b> |
|--------------------|---------------------------|-----------------------------|----------------|-------------------------------|
| B1*                | 1.2 mg                    | 1.1 mg                      | 0.2 to 0.3 mg  | 0.5 to 1.2 mg                 |
| B2*                | 1.3 mg                    | 1.1 mg                      | 0.3 to 0.4 mg  | 0.5 to 1.3 mg                 |
| Niacin*            | 16 mg                     | 14 mg                       | 2 to 4 mg      | 6 to 16 mg                    |

| <b>Vitamin 70)</b> | <b>Men (age 23 to 70)</b> | <b>Women (age 23 to 70)</b> | <b>Infants</b> | <b>Children (age 1 to 18)</b> |
|--------------------|---------------------------|-----------------------------|----------------|-------------------------------|
| B6                 | 1.3 mg                    | 1.3 mg                      | 0.1 to 0.3 mg  | 0.5 to 1.3 mg                 |
| B12                | 2.4 µg                    | 2.4 µg                      | 0.4 to 0.5 µg  | 0.9 to 2.4 µg                 |

\*Requirements per 1,000 kcal of dietary intake.

## Causes and Incidence

Thiamine deficiency results from malabsorption or inadequate dietary intake of vitamin B<sub>1</sub>. It also results from alcoholism, prolonged diarrhea, or from increased requirement, which can occur in pregnancy, lactation, and hyperthyroidism. Beriberi, a serious thiamine deficiency disease, is most prevalent in Asians, who subsist mainly on diets of unenriched rice and wheat. Although this disease is uncommon in the United States, alcoholics may develop cardiac (wet) beriberi with high-output heart failure, neuropathy, and cerebral disturbances. In times of stress (e.g., pregnancy), malnourished young adults may develop beriberi; infantile beriberi may appear in infants on low-protein diets or in those breast-fed by thiamine-deficient mothers.

Riboflavin deficiency (ariboflavinosis) results from a diet deficient in milk, meat, fish, legumes, and green, leafy vegetables. Alcoholism or prolonged diarrhea may also induce riboflavin deficiency. Exposure of milk to sunlight or treatment of legumes with baking soda can destroy riboflavin.

Niacin deficiency, in its advanced form, produces pellagra, which affects the skin, central nervous system (CNS), and GI tract. (See *Recognizing pellagra*, page 493.) Although this deficiency is now seldom found in the United States, it was once common among Southerners who subsisted mainly on corn and consumed minimal animal protein. (Corn is low in niacin and in available tryptophan, the amino acid from which the body synthesizes niacin.) Niacin deficiency is still common in parts of Egypt, Romania, Africa, Serbia, and Montenegro, where corn is the dominant staple food. Niacin deficiency can also occur secondary to carcinoid syndrome or Hartnup disease.

## Recognizing pellagra

This patient with pellagra shows dark, scaly, advanced dermatitis. In advanced niacin deficiency, such dermatitis usually occurs on areas exposed to the sun.



Pyridoxine deficiency usually results from destruction of pyridoxine in infant formulas by autoclaving. A frank deficiency is uncommon in adults, except in patients taking pyridoxine antagonists, such as isoniazid and penicillamine.

Cobalamin deficiency most commonly results from an absence of intrinsic factor in gastric secretions, or an absence of receptor sites after ileal resection. Other causes include malabsorption syndromes associated with sprue, intestinal worm infestation, regional ileitis, and gluten enteropathy, and a diet low in animal protein.

## Pathophysiology

Thiamine deficiency ( $B_1$ )—when healthy individuals are deprived of thiamine, thiamine stores are depleted within 1 month. However, within a week after thiamine intake stops, healthy people develop a resting tachycardia, weakness, and decreased deep tendon reflexes; some people develop a peripheral neuropathy.

Riboflavin deficiency ( $B_2$ )—riboflavin deficiency can alter iron absorption and cause an anemia that leads to fatigue. Riboflavin is involved in red blood cell (RBC) production and transportation of oxygen to the cells. Improving the amount of riboflavin in the body can increase circulating hemoglobin levels and increase red cell production. Collagen is a protein found in most skin and hair, so riboflavin is necessary to maintain a good collagen level. Taking supplements of riboflavin is also a cure for migraines. Research showed that 400 mg of riboflavin a day had demonstrated efficacy in prevention of a migraine in adults, but it must be taken for a minimum of 3 months for good results. This is most likely because mitochondrial dysfunction has been shown to play a role in migraines, and riboflavin is a precursor of flavin cofactors of the electron transport chain.

Niacin deficiency ( $B_3$ )—niacin is essential for adequate cellular function because of its required roles in two similar but distinct coenzymes (i.e., nicotinamide adenine dinucleotide [NAD] and nicotinamide adenine dinucleotide phosphate [NADP]). Both of these are cofactors that can be recycled by serving as both oxidizing (NAD, NADP) and reducing (NADH, NADPH) agents.

During the oxidation of glucose and other intermediary metabolites, a substantial amount of chemical energy is released. NAD/NADH are able to transfer electrons in a process that captures the energy by generating high-energy phosphate bonds. The synthesized ATP then provides the energy necessary for other reactions of intermediary metabolism that simultaneously regenerate NAD from the reduced NADH. A portion of this cofactor is also converted to NADP/NADPH, which plays several distinct roles. Reduced NADPH is used in reactions that detoxify reactive oxygen species, that metabolize drugs in a cytochrome P450 system, and that support lipid biosynthesis.

Pyridoxine deficiency ( $B_6$ )—after absorption, pyridoxine, pyridoxamine, and pyridoxal are transported into hepatic cells by facilitated diffusion. Pyridoxal kinase phosphorylates pyridoxine and pyridoxamine, after which

they are converted to pyridoxal 5'-phosphate (PLP) by a flavin-dependent enzyme. PLP either remains in the hepatocyte, where it is bound to an apoenzyme, or it is released into the serum, where it is tightly bound to albumin. Free pyridoxal is degraded by alkaline phosphatase, hepatic and renal aldehyde oxidases, and pyridoxal dehydrogenase.

Pyridoxine 5'-phosphate is an essential cofactor in various transamination, decarboxylation, and synthesis pathways involving carbohydrates, sphingolipids, sulfur-containing amino acids, heme, and neurotransmitters. PLP is a coenzyme of tryptophan, methionine, and gamma aminobutyric acid (GABA) metabolism. With methionine deficiency, *S*-adenosylmethionine accumulates, resulting in the inhibition of sphingolipid and myelin synthesis. Tryptophan is a precursor to several neurotransmitters and is required for niacin production. Thus, pyridoxine deficiency can cause a syndrome indistinguishable from pellagra. PLP is a cofactor for glutamic acid decarboxylase, the enzyme that produces GABA, such that PLP deficiency results in insufficient GABA. Since GABA is the major inhibitor cortical neurotransmitter, PLP deficiency can lead to seizures. Interestingly, pyridoxine-dependent seizures are not caused by a pyridoxine deficiency per se but rather due to an increased depletion of PLP.

The neurotransmitters dopamine, serotonin, epinephrine, norepinephrine, glycine, glutamate, and GABA also require PLP for their production. Homocysteine metabolism is dependent on pyridoxine, and high homocysteine levels can result from pyridoxine deficiency.

Cobalamin deficiency ( $B_{12}$ )—cobalamin deficiency is caused by the failure of gastric parietal cells to produce sufficient IF (a gastric protein secreted by parietal cells) to permit the absorption of adequate quantities of dietary vitamin  $B_{12}$ . Other disorders that interfere with the absorption and metabolism of vitamin  $B_{12}$  can produce cobalamin deficiency, with the development of a macrocytic anemia and neurologic complications.

## Complications

- ◆ heart failure
- ◆ neuropathy
- ◆ pellagra
- ◆ anemia

- ◆ seizures

## Signs and Symptoms

Thiamine deficiency causes polyneuritis and, possibly, Wernicke encephalopathy and Korsakoff psychosis. In infants (infantile beriberi), this deficiency produces edema, irritability, abdominal pain, pallor, vomiting, loss of voice and, possibly, seizures. In wet beriberi, severe edema starts in the legs and moves up through the body; dry beriberi causes multiple neurologic symptoms and an emaciated appearance. Thiamine deficiency may also cause cardiomegaly, palpitations, tachycardia, dyspnea, and circulatory collapse. Constipation and indigestion are common; ataxia, nystagmus, and ophthalmoplegia are also possible.

Riboflavin deficiency characteristically causes cheilosis (cracking of the lips and corners of the mouth), sore throat, and glossitis. It may also cause seborrheic dermatitis in the nasolabial folds, scrotum, and vulva and, possibly, generalized dermatitis involving the arms, legs, and trunk. This deficiency can also affect the eyes, producing burning, itching, light sensitivity, tearing, and vascularization of the corneas. Late-stage riboflavin deficiency causes neuropathy, mild anemia and, in children, growth retardation.

Niacin deficiency in its early stages produces fatigue, anorexia, muscle weakness, headache, indigestion, mild skin eruptions, weight loss, and backache. In advanced stages (pellagra), it produces dark, scaly dermatitis, especially on exposed body parts, that makes the patient appear to be severely sunburned. The mouth, tongue, and lips become red and sore, which may interfere with eating. Common GI symptoms include nausea, vomiting, and diarrhea. Associated CNS aberrations—confusion, disorientation, and neuritis—may become severe enough to induce hallucinations and paranoia. Because of this triad of symptoms, pellagra is sometimes called a “3-D” syndrome—dementia, dermatitis, and diarrhea. If not reversed by therapeutic doses of niacin, pellagra can be fatal.

Pyridoxine deficiency in infants causes a wide range of symptoms: dermatitis, occasional cheilosis or glossitis unresponsive to riboflavin therapy, abdominal pain, vomiting, ataxia, and seizures. This deficiency can also lead to CNS disturbances.

Cobalamin deficiency causes pernicious anemia, which produces anorexia, weight loss, abdominal discomfort, constipation, diarrhea, and

glossitis; peripheral neuropathy; and, possibly, ataxia, spasticity, and hyperreflexia.

## Diagnosis

The following values confirm vitamin B deficiency.

- ◆ *Thiamine deficiency*—commonly measured as micrograms per deciliter in a 24-hour urine collection. Deficiency levels are age-related: 1 to 3 years, less than 120; 4 to 6 years, less than 85; 7 to 9 years, less than 70; 10 to 12 years, less than 60; 13 to 15 years, less than 50; adults, less than 27; pregnant women, less than 23 (second trimester), less than 21 (third trimester).
- ◆ *Riboflavin deficiency*—measured as micrograms per gram of creatinine in a 24-hour urine collection. Deficiency levels are age-related: 1 to 3 years, less than 150; 4 to 6 years, less than 100; 7 to 9 years, less than 85; 10 to 15 years, less than 70; adults, less than 27; pregnant women, less than 39 (second trimester), less than 30 (third trimester).
- ◆ *Niacin deficiency*—measured by *n*-methyl nicotinamide in a 24-hour urine collection as micrograms per gram of creatinine. Deficiency levels in adults are less than 0.5; in pregnant women, less than 0.5 (first trimester), less than 0.6 (second trimester), and less than 0.8 (third trimester).
- ◆ *Pyridoxine deficiency*—xanthurenic acid more than 50 mg/day in 24-hour urine collection after administration of 10 g of L-tryptophan; decreased levels of serum and RBC transaminases; reduced excretion of pyridoxic acid in urine.
- ◆ *Cobalamin deficiency*—cobalamin serum levels less than 170 pg/mL. Tests to discover the deficiency's cause include gastric analysis and hemoglobin studies. In addition, the Schilling test measures absorption of radioactive cobalamin with and without intrinsic factor; however, it is rarely used.

## Treatment

Diet and supplementary vitamins can correct or prevent vitamin B deficiencies, as follows.

- ◆ *Thiamine deficiency*—a high-protein diet, with adequate calorie intake, possibly supplemented by B complex vitamins for early symptoms.

Thiamine-rich foods include pork, peas, wheat bran, oatmeal, and liver. Alcoholic beriberi may require thiamine supplements or thiamine hydrochloride as part of a B complex concentrate.

- ◆ *Riboflavin deficiency*—supplemental riboflavin in patients with intractable diarrhea or increased need for riboflavin related to growth, pregnancy, lactation, or wound healing. Good sources of riboflavin are meats, enriched flour, milk and dairy products, eggs, cereal, and green, leafy vegetables. Acute riboflavin deficiency requires daily oral doses of riboflavin alone or with other B complex vitamins. Riboflavin phosphate can also be administered I.V. or I.M.
- ◆ *Niacin deficiency*—supplemental B complex vitamins and dietary enrichment in patients at risk because of marginal diets or alcoholism. Meats, fish, peanuts, brewer's yeast, enriched breads, and cereals are rich in niacin; milk and eggs, in tryptophan. Confirmed niacin deficiency requires daily doses of niacinamide orally or I.V.
- ◆ *Pyridoxine deficiency*—prophylactic pyridoxine therapy in infants and in children with a seizure disorder; supplemental B complex vitamins in patients with anorexia, malabsorption, or those taking isoniazid or penicillamine. Some women who take hormonal contraceptives may have to supplement their diets with pyridoxine. Confirmed pyridoxine deficiencies require oral or parenteral pyridoxine. Children with seizures stemming from metabolic dysfunction may require daily doses of 200 to 600 mg pyridoxine.
- ◆ *Cobalamin deficiency*—parenteral cobalamin in patients with reduced gastric secretion of hydrochloric acid, lack of intrinsic factor, some malabsorption syndromes, or ileum resections. Strict vegetarians may have to supplement their diets with oral vitamin B<sub>12</sub>. Depending on the deficiency's severity, supplementary cyanocobalamin or methylcobalamin is usually given parenterally for 5 to 10 days, followed by monthly or daily vitamin B<sub>12</sub> supplements.

## Special Considerations

An accurate dietary history provides a baseline for effective dietary counseling.

- ◆ Identify and observe patients who are at risk for vitamin B deficiencies —alcoholics, the elderly, pregnant women, oral hormonal contraceptive

users (vitamins B<sub>6</sub> and B<sub>12</sub>), and people on limited diets.

- ◆ Administer prescribed supplements. Make sure patients understand how important it is that they adhere strictly to their prescribed treatment for the rest of their lives. Watch for adverse effects from large doses of niacinamide, such as a flushed sensation or hot flashes, in patients with niacin deficiency. Remember, prolonged intake of niacin can cause hepatic dysfunction. Caution patients with Parkinson disease receiving pyridoxine that this drug can impair response to levodopa therapy.
- ◆ Explain all tests and procedures. Reassure patients that, with treatment, the prognosis is good. Refer patients to appropriate assistance agencies if their diets are inadequate due to adverse socioeconomic conditions.



### PREVENTION

- ◆ *Encourage the patient to follow a well-balanced diet.*
- ◆ *Vitamin B<sub>12</sub> injections can prevent anemia after surgeries known to cause vitamin B<sub>12</sub> deficiency.*

## VITAMIN C DEFICIENCY

Vitamin C (ascorbic acid) deficiency leads to scurvy or inadequate production of collagen, an extracellular substance that binds the cells of the teeth, bones, and capillaries. It's essential for wound healing and burn recovery. Vitamin C is also an important factor in metabolizing such amino acids as tyrosine and phenylalanine. It also acts as a reductant, activating enzymes in the body, as well as converting folic acid into useful components.

Severe vitamin C deficiency results in scurvy, evidenced by hemorrhagic tendencies and abnormal osteoid and dentin formation.

### Causes and Incidence

This deficiency's primary cause is a diet lacking in vitamin C-rich foods, such as citrus fruits, tomatoes, cabbage, broccoli, spinach, and berries. Because the body can't store this water-soluble vitamin in large amounts, the supply needs to be replenished daily. Other causes include:

- ◆ destruction of vitamin C in foods by overexposure to air or by overcooking

- ◆ excessive ingestion of vitamin C during pregnancy, which causes the neonate to require large amounts of the vitamin after birth
- ◆ marginal intake of vitamin C during periods of physiologic stress—caused by infectious disease, for example—which can deplete tissue saturation of vitamin C

Historically common among sailors and others deprived of fresh fruits and vegetables for long periods of time, vitamin C deficiency is uncommon today in the United States, except in alcoholics, people on restricted-residue diets, and infants weaned from breast milk to cow's milk without a vitamin C supplement.

## Pathophysiology

Humans, other primates, and guinea pigs are unable to synthesize L-ascorbic acid (vitamin C); therefore, they require it in their diet. The enzyme L-gulonolactone oxidase, which would usually catalyze the conversion of L-gluconogammalactone to L-ascorbic acid, is defective due to a mutation or inborn error in carbohydrate metabolism.

Ascorbic acid is metabolized in the liver by oxidation and sulfation. The renal threshold for excretion by the kidney in urine is approximately 1.4 mg/100 mL plasma. Excess amounts of ascorbic acid are excreted unchanged or as metabolites. When body tissue or plasma concentrations of vitamin C are low, excretion of the vitamin is decreased. Scurvy occurs after vitamin C has been eliminated from the diet for at least 3 months and when the body pool falls below 350 mg.

## Complication

- ◆ Scurvy

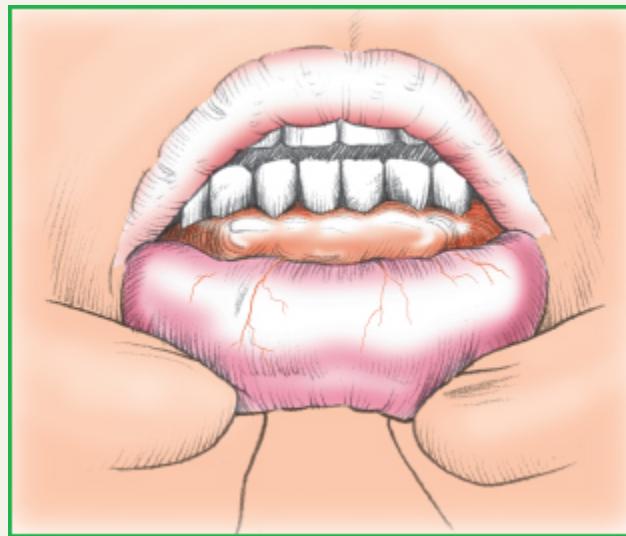
## Signs and Symptoms

Clinical features of vitamin C deficiency appear as capillaries become increasingly fragile. In an adult, it produces petechiae, ecchymoses, follicular hyperkeratosis (especially on the buttocks and legs), anemia, anorexia, limb and joint pain (especially in the knees), pallor, weakness, swollen or bleeding gums, loose teeth, lethargy, insomnia, poor wound healing, and ocular hemorrhages in the bulbar conjunctivae. (See *Scurvy's effect on gums and legs.*) Vitamin C deficiency can also cause beading,

fractures of the costochondral junctions of the ribs or epiphysis, and such psychological disturbances as irritability, depression, hysteria, and hypochondriasis.

### **Scurvy's effect on gums and legs**

In adults, scurvy causes swollen or bleeding gums and loose teeth.



It also causes follicular hyperkeratosis, usually on the legs.



In a child, vitamin C deficiency produces tender, painful swelling in the legs, causing the child to lie with the legs partially flexed. Other symptoms include fever, diarrhea, and vomiting.

## Diagnosis

**Dx CONFIRMING DIAGNOSIS** Serum ascorbic acid levels less than 0.2 mg/dL and white blood cell ascorbic acid levels less than 30 mg/dL help confirm the diagnosis.

Dietary history revealing an inadequate intake of ascorbic acid suggests vitamin C deficiency. A capillary fragility test may be performed on the patient's forearm with a blood pressure cuff; it's positive if more than 10 petechiae form after 5 minutes of pressure.

## Treatment

Because scurvy may be fatal, treatment begins immediately to restore adequate vitamin C intake with daily doses of 100 to 200 mg synthetic vitamin C or orange juice in mild disease and with doses as high as 500 mg/day in severe disease. Symptoms usually subside in 2 to 3 days; hemorrhages and bone disorders, in 2 to 3 weeks.

## Special Considerations

- ◆ Administer ascorbic acid orally or by slow I.V. infusion, as indicated. Avoid moving the patient unnecessarily to avoid irritating painful joints and muscles. Encourage the patient to drink orange juice.
- ◆ Explain the importance of supplemental ascorbic acid. Counsel the patient and family about good dietary sources of vitamin C.
- ◆ Advise against taking too much vitamin C. Explain that excessive doses of ascorbic acid may cause nausea, diarrhea, and renal calculi formation and may also interfere with anticoagulant therapy.



**PREVENTION** Patients unable or unwilling to consume foods rich in vitamin C or those facing surgery should take daily supplements of ascorbic acid. The recommended daily allowance is 75 to 90 mg/day. Vitamin C supplementation may also prevent this deficiency in recently weaned infants or those drinking formula not fortified with vitamin C.

## VITAMIN D DEFICIENCY

Vitamin D deficiency, commonly called *rickets*, causes failure of normal bone calcification, which occurs through several mechanisms: decreased calcium and phosphorus (the major components of bone) from the intestines, increased excretion of calcium from renal tubules, and increased parathyroid secretion resulting in increased release of calcium from the bone. The deficiency results in rickets in infants and young children and osteomalacia in adults. With treatment, the prognosis is good. However, in rickets, bone deformities usually persist, whereas in osteomalacia, such deformities may disappear.

## Causes and Incidence

Vitamin D deficiency results from inadequate dietary intake of preformed vitamin D, malabsorption of vitamin D, or too little exposure to sunlight.

Once a common childhood disease, rickets is now rare in the United States but occasionally appears in breast-fed infants who don't receive a vitamin D supplement or in infants receiving a formula with a nonfortified milk base. This deficiency may also occur in overcrowded urban areas in which smog limits sunlight penetration. Incidence is highest in black children who, because of their skin color, absorb less sunlight. (Solar ultraviolet rays irradiate 7-dehydrocholesterol, a precursor of vitamin D, to form calciferol.)

Osteomalacia, also uncommon in the United States, is most prevalent in Asia, among young multiparas who eat a cereal diet and have minimal exposure to sunlight. Other causes include:

- ◆ vitamin D-resistant rickets (refractory rickets, familial hypophosphatemia) from an inherited impairment of renal tubular reabsorption of phosphate (from vitamin D insensitivity)
- ◆ conditions that lower absorption of fat-soluble vitamin D, such as chronic pancreatitis, celiac disease, Crohn disease, cystic fibrosis, gastric or small-bowel resections, fistulas, colitis, and biliary obstruction
- ◆ hepatic or renal disease, which interferes with the formation of hydroxylated calciferol, necessary to initiate the formation of a calcium-binding protein in intestinal absorption sites
- ◆ malfunctioning parathyroid gland (decreased secretion of parathyroid hormone [PTH]), which contributes to calcium deficiency (normally, vitamin D controls calcium and phosphorus absorption through the intestine) and interferes with activation of vitamin D in the kidneys

## Pathophysiology

Inadequate circulation of 25(OH)D is associated with elevated PTH; this condition is called secondary hyperparathyroidism. The rise in PTH may result in increased mobilization of calcium from the bone, which leads to decreased mineralization of the bone.

Of note, prolonged exposure to the sun does not cause vitamin D toxicity. This is because after prolonged UVB radiation exposure, the vitamin D made in the skin is further degraded to the inactive vitamin D metabolites tachysterol and lumisterol.

## Complications

- ◆ Chronic skeletal pain
- ◆ Skeletal deformities
- ◆ Skeletal fractures

## Signs and Symptoms

Early indications of vitamin D deficiency are profuse sweating, restlessness, and irritability. Chronic deficiency induces numerous bone malformations due to softening of the bones: bowlegs, knock-knees, rachitic rosary (beading of ends of ribs), enlargement of wrists and ankles, pigeon breast, delayed closing of the fontanels, softening of the skull, and bulging of the forehead. (See *Recognizing bowlegs*.)

### Recognizing bowlegs

This infant with rickets shows characteristic bowing of the legs.



Other rachitic features are poorly developed muscles (potbelly) and infantile tetany. Bone deformities may cause difficulty in walking and in climbing stairs, spontaneous multiple fractures, and lower back and leg pain.

## Diagnosis

Physical examination, dietary history, and laboratory tests establish the diagnosis. Test results that suggest vitamin D deficiency include plasma calcium serum levels less than 7.5 mg/dL, serum inorganic phosphorus levels less than 3 mg/dL, serum citrate levels less than 2.5 mg/dL, and alkaline phosphatase levels less than 4 Bodansky U/dL.



**CONFIRMING DIAGNOSIS** *X-rays confirm the diagnosis by showing characteristic bone deformities and abnormalities such as Looser zones (pseudofractures).*

## Treatment

For osteomalacia and rickets—except when caused by malabsorption—treatment consists of oral doses of vitamin D or sources such as fish, liver, and processed milk. Exposure to sunlight is encouraged. For rickets refractory to vitamin D or in rickets accompanied by hepatic or renal disease, treatment includes 25-hydroxycholecalciferol, 1,25-dihydroxycholecalciferol, or a synthetic analog of active vitamin D. Replacement of deficient calcium and phosphorus also helps to eliminate most symptoms of rickets. Positioning or bracing may be used to reduce or prevent deformities; some skeletal deformities may require corrective surgery.

## Special Considerations

- ◆ Obtain a dietary history to assess the patient's current vitamin D intake. Encourage the patient to eat foods high in vitamin D—fortified milk, fish liver oils, herring, liver, and egg yolks—and get sufficient sun exposure. If deficiency is due to socioeconomic conditions, refer the patient to appropriate community agencies.

- ◆ If the patient must take vitamin D for a prolonged period, tell the patient to watch for signs of vitamin D toxicity (headache, nausea, constipation and, after prolonged use, renal calculi).



## PREVENTION

- ◆ *Administer supplementary aqueous preparations of vitamin D for chronic fat malabsorption, hydroxylated cholecalciferol for refractory rickets, and supplemental vitamin D for breast-fed infants.*
- ◆ *Consider genetic counseling for a patient with a family history of inherited disorders that can cause rickets.*

## VITAMIN E DEFICIENCY

Vitamin E (tocopherol) appears to act primarily as an antioxidant, preventing intracellular oxidation of polyunsaturated fatty acids and other lipids. It protects body tissue from damage caused by unstable substances called *free radicals*, which can harm cells, tissues, and organs and are believed to be one of the causes of aging's degenerative process. Vitamin E is also important in the formation of RBCs and helps the body to use vitamin K. Vitamin E deficiency usually manifests as hemolytic anemia in low-birth-weight or premature neonates. With treatment, prognosis is good.

### Causes and Incidence

Vitamin E deficiency in infants usually results from consuming formulas high in polyunsaturated fatty acids that are fortified with iron but not vitamin E. Such formulas increase the need for antioxidant vitamin E because the iron supplement catalyzes the oxidation of RBC lipids. A neonate has low tissue concentrations of vitamin E to begin with because only a small amount passes through the placenta; the mother retains most of it. Because vitamin E is a fat-soluble vitamin, deficiency develops in conditions associated with fat malabsorption, such as kwashiorkor, celiac disease, or cystic fibrosis. These conditions may induce megaloblastic or hemolytic anemia and creatinuria, all of which are reversible with vitamin E administration.

Vitamin E deficiency is uncommon in adults but is possible in people whose diets are high in polyunsaturated fatty acids, which increase vitamin

E requirements, and in people with vitamin E malabsorption, which impairs RBC survival.

## Complication

- ◆ Hemolytic anemia

## Signs and Symptoms

Vitamin E deficiency is difficult to recognize, but its early symptoms include edema and skin lesions in infants and muscle weakness or intermittent claudication in adults. In premature neonates, vitamin E deficiency produces hemolytic anemia, thrombocytopenia, and erythematous papular skin eruption, followed by desquamation.

## Diagnosis

 **CONFIRMING DIAGNOSIS** *Dietary and medical histories suggest vitamin E deficiency. Serum alpha-tocopherol levels below 0.5 mg/dL in adults and below 0.2 mg/dL in infants confirm it. Creatinuria, increased creatine kinase levels, hemolytic anemia, and an elevated platelet count generally support the diagnosis.*

## Treatment

Replacement of vitamin E with a water-soluble supplement, either oral or parenteral, is the only appropriate treatment.

## Special Considerations

- ◆ As indicated, prevent deficiency by providing vitamin E supplements for low-birth-weight neonates receiving formulas not fortified with vitamin E and for adults with vitamin E malabsorption. Many commercial multivitamin supplements are easily absorbed by patients with vitamin E malabsorption.
- ◆ If vitamin E deficiency is related to socioeconomic conditions, refer the patient to appropriate community agencies.



## PREVENTION

- *Inform new mothers who plan to breast-feed that human milk provides adequate vitamin E.*
- *Encourage adult patients to eat foods high in vitamin E; good sources include vegetable oils (corn, safflower, soybean, cottonseed); whole grains; dark green, leafy vegetables; nuts; and legumes. Tell them that heavy consumption of polyunsaturated fatty acids increases the need for vitamin E.*

## VITAMIN K DEFICIENCY

Deficiency of vitamin K, an element necessary for formation of prothrombin and other clotting factors in the liver, produces abnormal bleeding. If the deficiency is corrected, the prognosis is excellent.

### Causes and Incidence

Vitamin K deficiency is common among neonates in the first few days postpartum due to poor placental transfer of vitamin K and inadequate production of vitamin K-producing intestinal flora. Its other causes include prolonged use of drugs, such as the anticoagulant warfarin and antibiotics that destroy normal intestinal bacteria; decreased flow of bile to the small intestine from obstruction of the bile duct or bile fistula; malabsorption of vitamin K due to sprue, pellagra, bowel resection, ileitis, or ulcerative colitis; chronic hepatic disease, with impaired response of hepatic ribosomes to vitamin K; and cystic fibrosis, with fat malabsorption. Vitamin K deficiency seldom results from insufficient dietary intake of this vitamin.

### Pathophysiology

Vitamin K is necessary for the formation of prothrombin and other blood-clotting factors in the liver, and it also plays a role in bone metabolism. A form of the vitamin is produced by bacteria in the colon and can be utilized to some degree. Vitamin K deficiency causes impaired clotting of the blood and internal bleeding, even without injury.

### Complications

- ◆ Hemorrhagic disease
- ◆ Osteoporosis

## Signs and Symptoms

The cardinal sign of vitamin K deficiency is an abnormal bleeding tendency, accompanied by prolonged prothrombin time (PT); these signs disappear with vitamin K administration. Without treatment, bleeding may be severe and, possibly, fatal.

## Diagnosis

 **CONFIRMING DIAGNOSIS** *A PT that's 25% longer than the normal range of 10 to 20 seconds, measured by the Quick method, confirms the diagnosis of vitamin K deficiency after other causes of prolonged PT (such as anticoagulant therapy or hepatic disease) have been ruled out. The international normalized ratio (normal value, 0.8 to 1.2) is the more common method of assessing PT adequacy.*

Repetition of testing in 24 hours (and regularly during treatment) monitors the therapy's effectiveness.

## Treatment

Administration of vitamin K I.V. or I.M. corrects abnormal bleeding tendencies.

## Special Considerations



### PREVENTION

- Administer vitamin K to neonates and patients with fat malabsorption or with prolonged diarrhea from colitis, ileitis, or long-term antibiotic drug therapy.
- If the deficiency has a dietary cause, help the patient and family plan a diet that includes important sources of vitamin K, such as cauliflower, tomatoes, cheese, egg yolks, liver, and green, leafy vegetables.
- Warn against self-medication with or overuse of antibiotics, because these drugs destroy the intestinal bacteria needed to generate significant amounts of vitamin K.

## HYPERVITAMINOSES A AND D

Hypervitaminosis A is excessive accumulation of vitamin A; hypervitaminosis D, of vitamin D. Although these are toxic conditions, they usually respond well to treatment. They're most prevalent in infants and children, usually as a result of accidental or misguided overdosage by parents. A related, benign condition called *hypercarotenemia* results from excessive consumption of carotene, a chemical precursor of vitamin A.

## Causes and Incidence

Vitamins A and D are fat-soluble vitamins that accumulate in the body because they aren't dissolved and excreted in the urine. (See *Important facts about vitamins A and D*.) In most cases, hypervitaminoses A and D result from ingestion of excessive amounts of supplemental vitamin preparations. A single dose of more than 1 million units of vitamin A can cause acute toxicity; daily doses of 15,000 to 25,000 U taken over weeks or months have proven toxic in infants and children. For the same dose to produce toxicity in adults, ingestion over years is necessary. Doses of 100,000 IU of vitamin D daily for several months can cause toxicity in adults. Individuals who are at risk include those with hyperparathyroidism, kidney disease, sarcoidosis, tuberculosis, or histoplasmosis.

## Important facts about vitamins A and D

This table illustrates good sources of vitamins A and D, their recommended daily allowances, and the actions they produce.

| Vitamin Sources  | Recommended dietary allowance  | Action   |
|--|--|--|
| Vitamin A<br>◆ Carrots; sweet potatoes; dark green, leafy vegetables; butter; margarine; liver; egg yolk | ◆ Ages 1 to 13: 1,000 to 2,000 IU<br>◆ Ages ≥14: 2,300 to 3,000 IU<br>◆ Lactating women: 4,000 to 4,300 IU | ◆ Produces retinal pigment and maintains epithelial tissue |

| Vitamin Sources   | Recommended dietary allowance | Action   |
|---|-------------------------------|--|
| Vitamin D<br>Ultraviolet light; fortified foods (especially milk) | 600 IU daily                  | Promotes absorption and regulates metabolism of calcium and phosphorus |

## Pathophysiology

Vitamin A—The bioavailability of retinol is generally more than 80%, whereas the bioavailability and bioconversion of carotenes (i.e., provitamin A) are lower. These may be affected by species, molecular linkage, amount of carotene, nutritional status, genetic factors, and other interactions.

While in general the body absorbs retinoids and vitamin A very efficiently, it lacks the mechanisms to destroy excessive loads. Thus, the possibility of toxicity exists unless intake is carefully regulated. It has been suggested that earlier estimates of daily human requirements of vitamin A be revised downward.

Vitamin D—The acute toxic dose for vitamin D has not been established. The chronic toxic dose is more than 50,000 IU/day in adults. In infants younger than 6 months, 1,000 IU/day may be considered unsafe. However, a wide variance in potential toxicity exists for vitamin D.



**PEDIATRIC TIP** *In infants, giving 40,000 IU daily of vitamin D can cause toxicity in 1 to 4 months.*

Hypervitaminosis A may occur in patients receiving pharmacologic doses of vitamin A for dermatologic disorders. Hypervitaminosis D may occur in patients receiving high doses of the vitamin as treatment for hypoparathyroidism, rickets, and the osteodystrophy of chronic renal failure, and in infants who consume fortified milk and cereals plus a vitamin supplement. Concentrations of vitamin A in common foods are generally too low to pose a danger of excessive intake. However, hypercarotenemia results from excessive consumption of vegetables high in carotene (a protovitamin that the body converts into vitamin A), such as carrots, sweet potatoes, and dark green, leafy vegetables.

## Complications

- ◆ Liver damage
- ◆ Osteoporosis
- ◆ Excess calcium buildup (which could cause kidney damage)

## Signs and Symptoms

Chronic hypervitaminosis A produces anorexia, irritability, headache, hair loss, malaise, itching, vertigo, bone pain, bone fragility, and dry, peeling skin. It may also cause hepatosplenomegaly and emotional lability. Acute toxicity may also produce transient hydrocephalus and vomiting. (Hypercarotenemia produces yellow or orange skin coloration.)

Hypervitaminosis D causes anorexia, headache, nausea, vomiting, weight loss, polyuria, and polydipsia. Because vitamin D promotes calcium absorption, severe toxicity can lead to hypercalcemia, including calcification of soft tissues, as in the heart, aorta, and renal tubules. Lethargy, confusion, and coma may accompany severe hypercalcemia.

## Diagnosis

A thorough patient history suggests hypervitaminosis A.

 **CONFIRMING DIAGNOSIS** *An elevated serum vitamin A level (over 90 µg/dL) confirms hypervitaminosis A.*

Patient history and an elevated serum calcium level (over 10.5 µg/dL) suggest hypervitaminosis D.

 **CONFIRMING DIAGNOSIS** *An elevated serum vitamin D level confirms hypervitaminosis D.*

In children, X-rays showing calcification of tendons, ligaments, and subperiosteal tissues support this diagnosis.

 **CONFIRMING DIAGNOSIS** *An elevated serum carotene level (over 250 µg/dL) confirms hypercarotenemia.*

## Treatment

Withholding vitamin supplements usually corrects hypervitaminosis A quickly and hypervitaminosis D gradually. Hypercalcemia may persist for weeks or months after the patient stops taking vitamin D. Treatment for severe hypervitaminosis D may include glucocorticoids to control hypercalcemia and prevent renal damage. In the acute stage, diuretics or other emergency measures for severe hypercalcemia may be necessary. Hypercarotenemia responds well to a diet free of high-carotene foods.

## Special Considerations

- ◆ Keep the patient comfortable, and reassure the patient that symptoms will subside after they stop taking the vitamin.
- ◆ Make sure the patient or the parents of a child with these conditions understand that vitamins aren't innocuous. Explain the hazards associated with excessive vitamin intake. Point out that vitamin A and D requirements can easily be met with a diet containing dark green, leafy vegetables; fruits; and fortified milk or milk products.



**PREVENTION** Monitor serum vitamin A levels in patients receiving doses above the recommended daily allowance and serum calcium levels in patients receiving pharmacologic doses of vitamin D.

## IODINE DEFICIENCY

Iodine deficiency is the absence of sufficient levels of iodine to satisfy daily metabolic requirements. Because the thyroid gland uses most of the body's iodine stores, iodine deficiency is apt to cause hypothyroidism and thyroid gland hypertrophy (endemic goiter). Other effects of deficiency range from dental caries to cretinism in neonates born to iodine-deficient mothers. Iodine deficiency is most common in pregnant or lactating women due to their exaggerated metabolic need for this element. Iodine deficiency responds readily to treatment with iodine supplements.

## Causes and Incidence

Iodine deficiency usually results from insufficient intake of dietary sources of iodine, such as iodized table salt, seafood, and dark green, leafy vegetables. (Normal iodine requirements range from 35 µg/day for infants to 150 µg/day for lactating women; the average adult needs 1 µg/kg of body

weight.) Iodine deficiency may also result from an increase in metabolic demands during pregnancy, lactation, and adolescence.

## Pathophysiology

Dietary iodine is taken up readily through the gut in the form of iodide. From the circulation, it is concentrated in the thyroid gland by means of an energy-dependent sodium iodide symporter. In the follicle cells of the thyroid gland, four atoms of iodine are incorporated into each molecule of thyroxine ( $T_4$ ) and three atoms into each molecule of triiodothyronine ( $T_3$ ).

## Complications

- ◆ Hypothyroidism
- ◆ Goiter

## Signs and Symptoms

Clinical features of iodine deficiency depend on the degree of hypothyroidism that develops (in addition to the development of a goiter). Mild deficiency may produce only mild, nonspecific symptoms, such as lassitude, fatigue, and loss of motivation. Severe deficiency usually generates the typically overt and unmistakable features of hypothyroidism: bradycardia; decreased pulse pressure and cardiac output; weakness; hoarseness; dry, flaky, inelastic skin; puffy face; thick tongue; delayed relaxation phase in deep tendon reflexes; poor memory; hearing loss; chills; anorexia; and nystagmus. In women, iodine deficiency may also cause menorrhagia and amenorrhea.

Cretinism—hypothyroidism that develops in utero or in early infancy—is characterized by failure to thrive, neonatal jaundice, and hypothermia. By age 3 to 6 months, the infant may display spastic diplegia and signs and symptoms similar to those seen in infants with Down syndrome.

## Diagnosis



**CONFIRMING DIAGNOSIS** Abnormal laboratory test results include low thyroxine ( $T_4$ ) levels with high radioactive iodine ( $^{131}I$ ) uptake, low 24-hour urine iodine levels, and high thyroid-stimulating hormone levels. Radioiodine uptake test traces  $^{131}I$  in the thyroid 24 hours after

*administration; triiodothyronine-resin or  $T_4$ -resin uptake test shows values 25% below normal.*

## **Treatment**

Severe iodine deficiency requires administration of iodine supplements (potassium iodide [SSKI]). Mild deficiency may be corrected by increasing iodine intake through the use of iodized table salt and consumption of iodine-rich foods (seafood and green, leafy vegetables).

## **Special Considerations**

- ◆ Administer SSKI preparation in milk or juice to reduce gastric irritation and mask its metallic taste. To prevent tooth discoloration, tell the patient to drink the solution through a straw. Store the solution in a light-resistant container.



### **PREVENTION**

- ◆ *Recommend the use of iodized salt and consumption of iodine-rich foods for high-risk patients—especially adolescents and pregnant or lactating women.*
- ◆ *Advise pregnant women that severe iodine deficiency may produce cretinism in neonates, and instruct them to watch for early signs of iodine deficiency, such as fatigue, lassitude, weakness, and decreased mental function.*

## **ZINC DEFICIENCY**

Zinc, an essential trace element that's present in the bones, teeth, hair, skin, testes, liver, and muscles, is also a vital component of many enzymes. The prognosis is good with correction of the deficiency.

## **Causes and Incidence**

Zinc deficiency usually results from excessive intake of foods (containing iron, calcium, vitamin D, and the fiber and phytates in cereals) that bind zinc to form insoluble chelates that prevent its absorption. Occasionally, it results from blood loss due to parasitism and low intake of foods containing zinc. Alcohol and corticosteroids increase renal excretion of zinc.

Zinc deficiency is most common in people from underdeveloped countries, especially in the Middle East. Children are most susceptible to this deficiency during periods of rapid growth.

## Pathophysiology

Zinc promotes synthesis of deoxyribonucleic acid, ribonucleic acid and, ultimately, protein, and maintains normal blood concentrations of vitamin A by mobilizing it from the liver.

## Complications

- ◆ Mental lethargy
- ◆ Decreased wound healing
- ◆ Impaired immune function

## Signs and Symptoms

Zinc deficiency produces hepatosplenomegaly, sparse hair growth, soft and misshapen nails, poor wound healing, anorexia, hypogeesthesia (decreased taste acuity), dysgeusia (unpleasant taste), hyposmia (decreased odor acuity), dysosmia (unpleasant odor in nasopharynx), severe iron deficiency anemia, bone deformities and, when chronic, hypogonadism, dwarfism, and hyperpigmentation.

## Diagnosis



**CONFIRMING DIAGNOSIS** Fasting serum zinc levels below 70 µg/dL

confirm zinc deficiency and indicate altered phosphate metabolism, imbalance between aerobic and anaerobic metabolism, and decreased pancreatic enzyme levels.

## Treatment

Treatment consists of correcting the deficiency's underlying cause and administering zinc supplements, as necessary.

## Special Considerations

- ◆ Advise the patient to take zinc supplements with milk or meals to prevent gastric distress and vomiting.



**PREVENTION** Encourage a balanced diet that includes seafood, oatmeal, bran, meat, eggs, nuts, and dry yeast and the correct use of calcium and iron supplements.

## OBESITY

Obesity is an excess of body fat, generally 20% above ideal body weight. The prognosis for correction of obesity is poor: Fewer than 30% of patients succeed in losing 20 lb (9 kg), and only half of these maintain the loss over a prolonged period.

### Causes and Incidence

Obesity results from excessive calorie intake and inadequate expenditure of energy. Rates of obesity are climbing, and the percentage of children and adolescents who are obese has doubled in the past 20 years.

### Pathophysiology

Theories to explain this condition include hypothalamic dysfunction of hunger and satiety centers, genetic predisposition, abnormal absorption of nutrients, and impaired action of GI and growth hormones and of hormonal regulators such as insulin. An inverse relationship between socioeconomic status and the prevalence of obesity has been documented, especially in women. Obesity in parents increases the probability of obesity in children, from genetic or environmental factors, such as activity levels and learned patterns of eating. Psychological factors, such as stress or emotional eating, may also contribute to obesity.

### Complications

- ◆ Respiratory difficulties
- ◆ Hypertension
- ◆ Cardiovascular disease
- ◆ Diabetes mellitus
- ◆ Renal disease
- ◆ Gallbladder disease
- ◆ Psychosocial difficulties
- ◆ Premature death

## **Signs and Symptoms**

Obesity is a problem of too much weight typically resulting in a body mass index (BMI) of 30 or higher. (See *Diagnostic aids*.)

## **Diagnostic Aids**

Weight categories, overweight and obesity, are determined by using a person's height and weight to calculate the BMI. *Overweight* is defined as a BMI between 25.0 and 29.9. *Obesity* is defined as a BMI of 30 or higher. Measurement of the thickness of subcutaneous fat folds with calipers provides an approximation of total body fat. Although this measurement is reliable and isn't subject to daily fluctuations, it has little meaning for the patient in monitoring subsequent weight loss.

## **Treatment**

Successful management of obesity must decrease the patient's daily calorie intake while increasing their activity level. Effective treatment must be based on a balanced, low-calorie diet that eliminates foods high in fat or sugar. Lifelong maintenance of these improved eating and exercise patterns is necessary to achieve long-term benefits.

The popular low-carbohydrate diets offer no long-term advantage; rapid early weight reduction is due to loss of water, not fat. These and other crash or fad diets have the overwhelming drawback that they don't teach the patient long-term modification of eating patterns and often lead to the "yo-yo syndrome"—episodes of repeated weight loss followed by weight gain. This can be more detrimental than the obesity itself because of the severe stress it can place on the body.

Total fasting is an effective method of rapid weight reduction but requires close monitoring and supervision to minimize risks of ketonemia, electrolyte imbalance, hypotension, and loss of lean body mass. Prolonged fasting and very-low-calorie diets have been associated with sudden death, possibly resulting from cardiac arrhythmias caused by electrolyte abnormalities. These methods also neglect patient re-education, which is necessary for long-term weight maintenance. The best way to lose weight is to do so slowly, losing 1 to 2 lb/week.

Treatment may also include behavior modification techniques, which promote fundamental changes in eating habits and activity patterns.

Food and Drug Administration (FDA)-approved medications for chronic weight loss include Alli (orlistat), Qsymia (phentermine and topiramate), Belviq (lorcaserin), Contrave (naltrexone/bupropion), and Saxenda (liraglutide). These drugs promote weight loss by decreasing the absorption of dietary fat. They need to be combined with healthy eating and physical activity to be most effective.

Obesity may be treated surgically with restriction or malabsorptive procedures. Those with a BMI greater than 40 or with a BMI between 35 and 39.9 along with a weight-related health problem may qualify for weight-loss surgery. Vertical banded gastroplasty, gastric banding, and the sleeve procedure represent restrictive surgical procedures that aid in weight loss by decreasing the volume of food that the stomach can accommodate. Gastric bypass with a Roux-en-Y procedure produces weight loss by both restricting stomach capacity and inducing malabsorption; food is rerouted to bypass part of the small intestine. Nutrition counseling before and after these procedures is recommended to enhance safe weight loss, educate the patient about proper diet advancement, and monitor for nutritional deficiencies. Psychological and social support are also beneficial before and after surgery.

## Special Considerations

- ◆ Obtain an accurate diet history to identify the patient's eating patterns and the importance of food to their lifestyle. Ask the patient to keep a careful record of what, where, and when they eat to help identify situations that normally provoke overeating.
- ◆ Explain the prescribed diet carefully, and encourage compliance to improve health status.
- ◆ To increase calorie expenditure, promote increased physical activity, including an exercise program. Recommend varying activity levels according to the patient's general condition and cardiovascular status.
- ◆ Teach the grossly obese patient the importance of good skin care to prevent breakdown in moist skin folds. Recommend the regular use of powder to keep skin dry.



- Teach parents to avoid overfeeding their infants and to familiarize themselves with actual nutritional needs and optimum growth rates. Discourage parents from using food to reward or console their children, from emphasizing the importance of “clean plates,” and from allowing eating to prevent hunger rather than to satisfy it.
- Encourage physical activity and exercise, especially in children and young adults, to establish lifelong patterns. Suggest low-calorie snacks such as raw vegetables.

## PROTEIN-CALORIE MALNUTRITION

One of the most prevalent and serious depletion disorders, protein-calorie malnutrition (PCM) occurs as marasmus (protein-calorie deficiency), characterized by growth failure and wasting, and as kwashiorkor (protein deficiency), characterized by tissue edema and damage. Both forms vary from mild to severe and may be fatal, depending on the accompanying stress (particularly sepsis or injury) and duration of deprivation. PCM increases the risk of death from pneumonia, chickenpox, or measles.

### Causes and Incidence

Both kwashiorkor (edematous PCM) and marasmus (nondematous PCM) are common in underdeveloped countries and in areas in which dietary amino acid content is insufficient to satisfy growth requirements. Kwashiorkor typically occurs at about age 1, after infants are weaned from breast milk to a protein-deficient diet of starchy gruels or sugar water, but it can develop at any time during the formative years. Marasmus affects infants ages 6 to 18 months as a result of breast-feeding failure, or a debilitating condition such as chronic diarrhea.

In industrialized countries, PCM may occur secondary to chronic metabolic disease that decreases protein and calorie intake or absorption, or trauma that increases protein and calorie requirements.



**ELDER TIP** In the United States, PCM is estimated to occur to some extent in 50% of elderly people in nursing homes.

Those who aren't allowed anything by mouth for an extended period are at high risk of developing PCM. Conditions that increase protein-calorie requirements include severe burns and injuries, systemic infections, and cancer (accounts for the largest group of hospitalized patients with PCM).

Conditions that cause defective utilization of nutrients include malabsorption syndrome, short-bowel syndrome, and Crohn disease.

## **Pathophysiology**

Marasmus is an insufficient energy intake to match the body's requirements. As a result, the body draws on its own stores, resulting in emaciation. In kwashiorkor, adequate carbohydrate consumption and decreased protein intake lead to decreased synthesis of visceral proteins. The resulting hypoalbuminemia contributes to extravascular fluid accumulation. Impaired synthesis of B-lipoprotein produces a fatty liver.

Protein-energy malnutrition also involves an inadequate intake of many essential nutrients.

## **Complications**

- ◆ Mental and physical disabilities
- ◆ Coma
- ◆ Shock

## **Signs and Symptoms**

Children with chronic PCM are small for their chronological age and tend to be physically inactive, mentally apathetic, and susceptible to frequent infections. Anorexia and diarrhea are common.

In acute PCM, children are small, gaunt, and emaciated, with no adipose tissue. Skin is dry and "baggy," and hair is sparse and dull brown or reddish-yellow. Temperature is low; pulse rate and respirations are slowed. Such children are weak, irritable, and usually hungry, although they may have anorexia, with nausea and vomiting.

Unlike marasmus, chronic kwashiorkor allows the patient to grow in height, but adipose tissue diminishes as fat metabolizes to meet energy demands. Edema often masks severe muscle wasting; dry, peeling skin and hepatomegaly are common. Patients with secondary PCM show signs similar to marasmus, primarily loss of adipose tissue and lean body mass, lethargy, and edema. Severe secondary PCM may cause loss of immunocompetence.

## **Diagnosis**

 **CONFIRMING DIAGNOSIS** *Clinical appearance, dietary history, and anthropometry confirm PCM. If the patient doesn't suffer from fluid retention, weight change over time is the best index of nutritional status.*

The following factors support the diagnosis:

- ◆ height and weight less than 80% of standard for the patient's age and sex, and below-normal arm circumference and triceps skinfold
- ◆ serum albumin level less than 2.8 g/dL (normal: 3.3 to 4.3 g/dL)
- ◆ urinary creatinine (24-hour) level used to show lean body mass status by relating creatinine excretion to height and ideal body weight, to yield CHI.

## Treatment

The aim of treatment is to provide sufficient proteins, calories, and other nutrients for nutritional rehabilitation and maintenance. When treating severe PCM, restoring fluid and electrolyte balance parentally is the initial concern. Nutrition provision should not be increased until laboratory values, such as electrolyte level, stabilize. A patient who shows normal absorption may receive enteral nutrition after anorexia has subsided. When possible, the preferred treatment is oral feeding. Foods are introduced slowly due to the risk of refeeding syndrome. Carbohydrates are given first to supply energy, and then high-quality protein foods, especially milk, and protein-calorie supplements are given. A patient who's unwilling or unable to eat may require supplementary feedings through a nasogastric (NG) tube or total parenteral nutrition (TPN), which is given through a central venous catheter because of its higher osmolality. Peripheral parenteral nutrition, which has a lower osmolality than TPN and can be given through a peripheral I.V. line, is an alternative to TPN, but it's given less commonly. Accompanying infection must also be treated, preferably with antibiotics that don't inhibit protein synthesis. Cautious realimentation is essential to prevent complications from overloading the compromised metabolic system.

## Special Considerations

- ◆ Encourage the patient with PCM to consume as much nutritious food and beverage as possible. (It's often helpful to "cheer the patient on" as

he or she eats.) Assist the patient with eating if necessary. Cooperate closely with the dietitian to monitor intake, and provide acceptable meals and snacks.

- If TPN is necessary, observe strict sterile technique when handling catheters, tubes, and solutions and during dressing changes.



## PREVENTION

- *Watch for PCM in patients who have been hospitalized for a prolonged period, have had no oral intake for several days, or have cachectic disease.*
- *To help eradicate PCM in developing countries, encourage prolonged breast-feeding, educate mothers about their children's needs, and provide supplementary foods, as needed.*



**ELDER TIP** *If the older patient is anorexic, consider asking family members and other visitors to bring in special foods from home that may improve the patient's appetite. In addition, encouraging the family to collaborate on feeding a dependent patient can help promote recovery, enhance feelings of well-being, and stimulate the patient to eat more.*

## Metabolic Disorders

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### GALACTOSEMIA

Galactosemia is any disorder of galactose metabolism. It produces symptoms ranging from cataracts and liver damage to mental retardation and occurs in two forms: classic galactosemia and galactokinase deficiency galactosemia. Although a galactose-free diet relieves most symptoms, galactosemia-induced mental impairment is irreversible; some residual vision impairment may also persist.

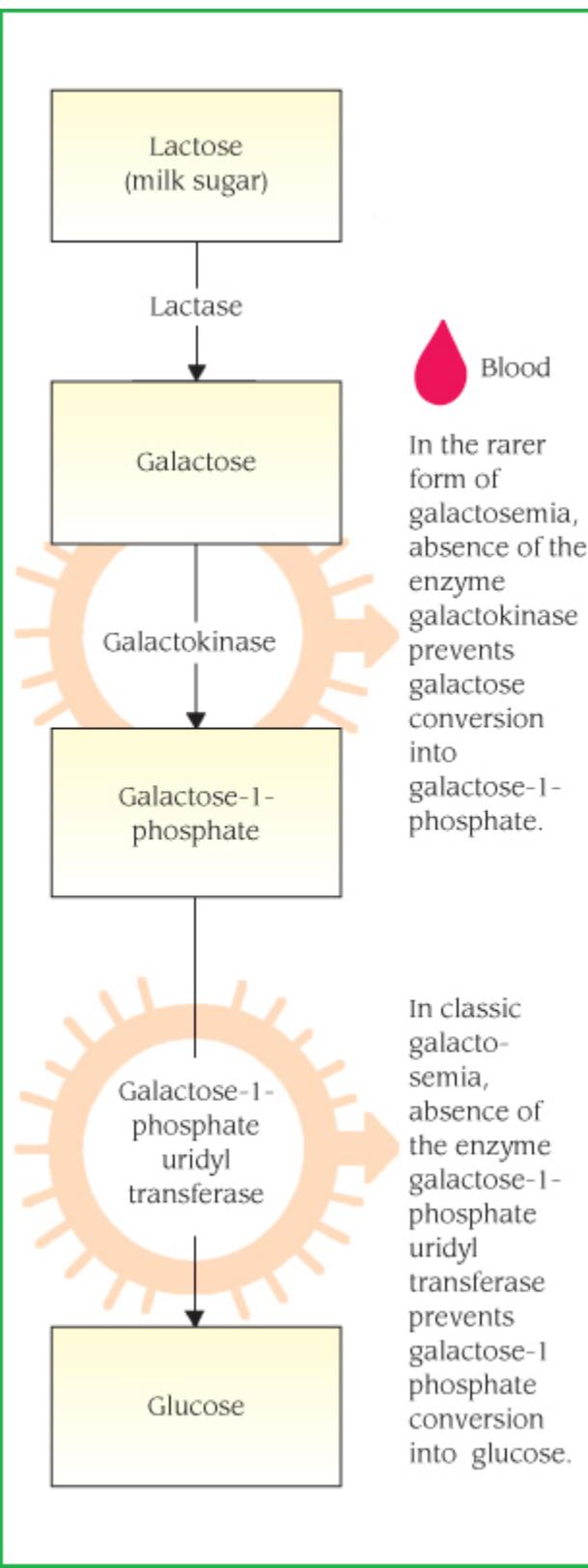
### Causes and Incidence

Both forms of galactosemia are inherited as autosomal recessive defects and occur in about 1 in 60,000 births in the United States. Up to 1.25% of the population is heterozygous for the classic galactosemia gene. Classic galactosemia results from a defect in the enzyme galactose-1-phosphate

uridyl transferase. (See *Metabolic pathway in galactosemia*.) Galactokinase deficiency galactosemia, the rarer form of this disorder, stems from a deficiency of the enzyme galactokinase. In both forms of galactosemia, the inability to normally metabolize the sugar galactose (which is mainly formed by digestion of the disaccharide lactose that's present in milk) causes galactose accumulation.

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### **Metabolic pathway in galactosemia**



## **Pathophysiology**

An elevated blood galactose concentration is the result of altered metabolism of galactose due to a genetic deficiency in enzyme activity or secondary hypergalactosemia due to liver disease (congenital hepatitis, patent ductus venosus, congenital hepatic arteriovenous malformation).

## **Complications**

- ◆ Ovarian dysfunction
- ◆ Cataracts
- ◆ Abnormal speech
- ◆ Cognitive impairment
- ◆ Motor delay
- ◆ Growth retardation

## **Signs and Symptoms**

In children who are homozygous for the classic galactosemia gene, signs are evident at birth or begin within a few days after milk ingestion, and include failure to thrive, vomiting, and diarrhea. Other clinical effects include liver damage (which causes jaundice, hepatomegaly, cirrhosis, and ascites), splenomegaly, galactosuria, proteinuria, and aminoaciduria. Cataracts may also be present at birth or develop later. Pseudotumor cerebri may occur.

Continued ingestion of galactose- or lactose-containing foods may cause mental retardation, malnourishment, progressive hepatic failure, and death—from the still-unknown process of galactose metabolites accumulating in body tissues. Although treatment may prevent mental impairment, galactosemia can produce a short attention span, difficulty with spatial and mathematical relationships, and apathetic, withdrawn behavior. Cataracts may be the only sign of galactokinase deficiency, resulting from the accumulation of galactitol, a metabolic by-product of galactose, in the lens.

## **Diagnosis**



**CONFIRMING DIAGNOSIS** Deficiency of the enzyme galactose-1-phosphate uridyl transferase in RBCs confirms classic galactosemia; decreased RBC levels of galactokinase confirm galactokinase deficiency.

*Prenatal diagnosis may be made by direct measurement of galactose-1-phosphate uridyl transferase.*

Related laboratory results include increased galactose levels in blood (normal value in children is <20 mg/dL) and urine (must use galactose oxidase to avoid confusion with other reducing sugars). Galactose measurements in blood and urine must be interpreted carefully because some children who consume large amounts of milk have elevated plasma galactose concentrations and galactosuria but aren't galactosemic. Also, neonates excrete galactose in their urine for about a week after birth; premature infants, even longer.

Other test results include:

- ◆ liver biopsy—typical acinar formation
- ◆ liver enzymes (aspartate aminotransferase, alanine aminotransferase levels)—elevated
- ◆ urinalysis—albumin in urine
- ◆ ophthalmoscopy—punctate lesions in the fetal lens nucleus (with treatment, cataracts regress)
- ◆ amniocentesis—prenatal diagnosis of galactosemia (recommended for heterozygous and homozygous parents)

## Treatment

Elimination of galactose and lactose from the diet causes most effects to subside. The infant is fed soy formula, meat-base formula, protein hydrolysate formula, or another lactose-free formula. The infant gains weight; liver anomalies, nausea, vomiting, galactosemia, proteinuria, and aminoaciduria disappear; and cataracts regress. As the child grows, a balanced, galactose-free diet must be maintained. Hormone replacement is sometimes necessary for puberty. A pregnant woman who's heterozygous or homozygous for galactosemia should also follow a galactose-restricted diet. Such a diet supports normal growth and development and may delay symptoms in the neonate.

## Special Considerations

- ◆ To eliminate galactose and lactose from an infant's diet, replace cow's milk formula or breast milk with a meat-base or soybean formula.

- ◆ Teach the parents about dietary restrictions and stress the importance of compliance. (See *Diet for galactosemia*.) Warn them to read medication labels carefully and avoid giving any medication that contains lactose fillers.

### **Diet for galactosemia**

A patient with galactosemia must follow a lactose-free diet. It's important for the patient to carefully read food labels to avoid milk and milk products, including dry milk products. The patient may eat these foods:

- ◆ fish and animal products (except brains and mussels)
- ◆ fresh fruits and vegetables (except peas and lima beans)
- ◆ only bread and rolls made from cracked wheat.

He should avoid these foods:

- ◆ dairy products
- ◆ puddings, cookies, cakes, pies
- ◆ food coloring
- ◆ instant potatoes
- ◆ canned and frozen foods (if lactose is listed as an ingredient)

- ◆ If the child has a learning disability, help parents secure educational assistance. Refer parents who want to have other children for genetic counseling. In some states, screening of all neonates for galactosemia is required by law.
- ◆ Instruct the parents to contact support groups, such as Parents of Galactosemic Children, for further information and support if appropriate.

## **GLYCOGEN STORAGE DISEASES**

Glycogen storage diseases consist of at least eight distinct errors of metabolism, all inherited, that alter the synthesis or degradation of glycogen, the form in which glucose is stored in the body. Normally, muscle and liver cells store glycogen. Muscle glycogen is used in muscle contraction; liver glycogen can be converted into free glucose, which can

then diffuse out of the liver cells to increase blood glucose levels. Glycogen storage diseases manifest as dysfunctions of the liver, heart, or musculoskeletal system. Symptoms vary from mild and easily controlled hypoglycemia to severe organ involvement that may lead to heart failure and respiratory failure.

## Causes and Incidence

Almost all glycogen storage diseases (types I through V and type VII) are transmitted as autosomal recessive traits. The transmission mode of type VI is unknown; type VIII may be an X-linked trait.

## Pathophysiology

The most common glycogen storage disease is type I—von Gierke, or hepatorenal glycogen storage disease—which results from a deficiency of the liver enzyme glucose-6-phosphatase. This enzyme converts glucose-6-phosphate into free glucose and is necessary for the release of stored glycogen and glucose into the bloodstream, to relieve hypoglycemia. Infants may die of acidosis before age 2; if they survive past this age, with proper treatment, they may grow normally and live to adulthood, with only minimal hepatomegaly. However, there's a danger of adenomatous liver nodules, which may be premalignant.

## Complications

- ◆ Heat intolerance
- ◆ Easy bruising
- ◆ Slowed growth
- ◆ Incomplete sexual development
- ◆ Hepatic adenomas
- ◆ Multiple organ failure

## Signs and Symptoms

Primary clinical features of the liver glycogen storage diseases (types I, III, IV, VI, and VIII) are hepatomegaly and rapid onset of hypoglycemia and ketosis when food is withheld. Symptoms of the muscle glycogen storage diseases (types II, V, and VII) include poor muscle tone; type II may result

in death from heart failure. (See *Rare forms of glycogen storage disease*, pages 508 and 509.)

## Rare forms of glycogen storage disease

| Type  | Clinical features  | Diagnostic test results   |
|---|--|---|
| <b>II (Pompe)</b>   |  |   |
| Absence of alpha-1,4-glucosidase (acid maltase)   | <ul style="list-style-type: none"> <li>◆ <i>Infants</i>: cardiomegaly, profound hypotonia and, occasionally, endocardial fibroelastosis (usually fatal before age 1 due to cardiac or respiratory failure)</li> <li>◆ <i>Some infants and young children</i>: muscle weakness and wasting, variable organ involvement (slower progression, usually fatal by age 19)</li> <li>◆ <i>Adults</i>: muscle weakness without organomegaly (slowly progressive but not fatal)</li> </ul> | <ul style="list-style-type: none"> <li>◆ <i>Muscle biopsy</i>: increased concentration of glycogen with normal structure; alpha-1,4-glucosidase deficiency</li> <li>◆ <i>Electrocardiogram (in infants)</i>: large QRS complexes in all leads; inverted T waves; shortened PR interval</li> <li>◆ <i>Electromyography (in adults)</i>: muscle fiber irritability; myotonic discharges</li> <li>◆ <i>Amniocentesis</i>: alpha-1,4-glucosidase deficiency</li> <li>◆ <i>Placenta or umbilical cord examination</i>: alpha-1,4-glucosidase deficiency</li> </ul> |
| <b>III (Cori)</b>   |  |   |
| Absence of debranching enzyme (amylo-1,6-glucosidase) (Note: predominant cause of glycogen storage disease in Israel) | <ul style="list-style-type: none"> <li>◆ <i>Young children</i>: massive hepatomegaly, which may disappear by puberty; growth retardation; moderate splenomegaly; hypoglycemia</li> <li>◆ <i>Adults</i>: progressive myopathy</li> <li>◆ Occasionally, moderate cardiomegaly, cirrhosis, muscle wasting, hypoglycemia</li> </ul>  | <ul style="list-style-type: none"> <li>◆ <i>Liver biopsy</i>: deficient debranching activity; increased glycogen concentration</li> <li>◆ <i>Laboratory tests (in children only)</i>: elevated aspartate aminotransferase or alanine aminotransferase levels; increased erythrocyte glycogen levels</li> </ul>  |
| <b>IV (Andersen)</b>  |  |   |
| Deficiency of branching enzyme (amylo-1,4-1,6-transglucosidase) (Note: extremely rare)                                | <ul style="list-style-type: none"> <li>◆ <i>Infants</i>: hepatosplenomegaly, ascites, muscle hypotonia; usually fatal before age 2 from progressive cirrhosis</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Liver biopsy</i>: deficient branching enzyme activity; glycogen molecule has longer outer branches</li> </ul>   |
| <b>V (McArdle)</b>  |  |   |

| Type   | Clinical features  | Diagnostic test results   |
|--|--|---|
| Deficiency of muscle phosphorylase           | <ul style="list-style-type: none"> <li>◆ <i>Children:</i> mild or no symptoms</li> <li>◆ <i>Adults:</i> muscle cramps and pain during strenuous exercise, possibly resulting in myoglobinuria and renal failure</li> <li>◆ <i>Older patients:</i> significant muscle weakness and wasting</li> </ul> | <ul style="list-style-type: none"> <li>◆ <i>Serum lactate:</i> no increase in venous levels in sample drawn from extremity after ischemic exercise</li> <li>◆ <i>Muscle biopsy:</i> lack of phosphorylase activity; increased glycogen content</li> </ul>   |
| <b>VI (Hers)</b>                             |  |   |
| Possible deficiency of hepatic phosphorylase | <ul style="list-style-type: none"> <li>◆ Mild symptoms (similar to those of type I), requiring no treatment</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Liver biopsy:</i> decreased phosphorylase b activity, increased glycogen concentration</li> </ul>   |
| <b>VII (Tarui)</b>                           |  |   |
| Deficiency of muscle phosphofructokinase     | <ul style="list-style-type: none"> <li>◆ Muscle cramps during strenuous exercise, resulting in myoglobinuria and possible renal failure</li> <li>◆ Reticulocytosis</li> </ul>  | <ul style="list-style-type: none"> <li>◆ <i>Serum lactate:</i> no increase in venous levels in sample drawn from extremity after ischemic exercise</li> <li>◆ <i>Muscle biopsy:</i> deficient phosphofructokinase; marked rise in glycogen concentration</li> <li>◆ <i>Blood studies:</i> low erythrocyte phosphofructokinase activity; reduced half-life of red blood cells</li> </ul> |
| <b>VIII</b>                                  |  |   |
| Deficiency of hepatic phosphorylase kinase   | <ul style="list-style-type: none"> <li>◆ Mild hepatomegaly</li> <li>◆ Mild hypoglycemia</li> </ul>   | <ul style="list-style-type: none"> <li>◆ <i>Liver biopsy:</i> deficient phosphorylase b kinase activity; increased glycogen concentration</li> <li>◆ <i>Blood study:</i> deficient phosphorylase b kinase in leukocytes</li> </ul>  |

In addition, type I may produce these symptoms:

- ◆ infants—acidosis, hyperlipidemia, GI bleeding, coma
- ◆ children—low resistance to infection and, without proper treatment, short stature
- ◆ adolescents—gouty arthritis and nephropathy; chronic tophaceous gout; bleeding (especially epistaxis); small superficial vessels visible in skin

due to impaired platelet function; fat deposits in cheeks, buttocks, and subcutaneous tissues; poor muscle tone; enlarged kidneys; xanthomas over extensor surfaces of arms and legs; steatorrhea; multiple, bilateral, yellow lesions in fundi; and osteoporosis, probably secondary to negative calcium balance. Correct treatment of glycogen storage disease should prevent all of these effects.

## Diagnosis

 **CONFIRMING DIAGNOSIS** Liver biopsy confirms the diagnosis by showing normal glycogen synthetase and phosphorylase enzyme activities but reduced or absent glucose-6-phosphatase activity. Glycogen structure is normal but amounts are elevated. Spectroscopy may be used to show abnormal muscle metabolism with the use of magnetic resonance imaging in specialized centers.

- ◆ Laboratory studies of plasma demonstrate low glucose levels but high levels of free fatty acids, triglycerides, cholesterol, and uric acid. Serum analysis reveals high pyruvic acid levels and high lactic acid levels. Prenatal diagnoses are available for types II, III, and IV.
- ◆ Injection of glucagon or epinephrine increases pyruvic and lactic acid levels but doesn't increase blood glucose levels. Glucose tolerance test curve typically shows depletional hypoglycemia and reduced insulin output. Intrauterine diagnosis is possible.

## Treatment

For type I, treatment aims to maintain glucose homeostasis and prevent secondary consequences of hypoglycemia through frequent feedings and constant nocturnal enteral feeding with Polycose or Vivonex. Treatment includes a low-fat diet, with normal amounts of protein and calories; carbohydrates should contain glucose or glucose polymers only. Physically modified cornstarch is also used to prevent hypoglycemia.

Therapy for type III includes frequent feedings (every 3 to 4 hours) and a high-protein diet. Type IV requires a high-protein, high-calorie diet; bed rest; diuretics; sodium restriction; and paracentesis, if necessary, to relieve ascites. Ultimately treat with liver transplant. Types V and VII require no

treatment except avoidance of strenuous exercise. No treatment is necessary for types VI and VIII; no effective treatment exists for type II.

## Special Considerations

When managing type I disease:

- ◆ Advise the patient or parents to include carbohydrate foods containing mainly starch in the diet and to sweeten foods with glucose only.
- ◆ Before discharge, teach the patient or family member how to use the enteral feeding equipment, monitor blood glucose levels with glucose reagent strips, and recognize symptoms of hypoglycemia.
- ◆ Watch for and report signs of infection (fever, chills, myalgia) and of hepatic encephalopathy (mental confusion, stupor, asterixis, coma) due to increased blood ammonia levels.

When managing other types, do the following.

- ◆ *Type II:* Explain test procedures, such as electromyography and electroencephalography (EEG), thoroughly.
- ◆ *Type III:* Instruct the patient to eat a high-protein diet (eggs, nuts, fish, meat, poultry, and cheese).
- ◆ *Type IV:* Watch for signs of hepatic failure (nausea, vomiting, irregular bowel function, clay-colored stools, right upper quadrant pain, jaundice, dehydration, electrolyte imbalance, edema, and changes in mental status, progressing to coma).

When caring for patients with types II, III, and IV glycogen storage disease, offer the patient and parents reassurance and emotional support. Recommend and arrange for genetic counseling, if appropriate.

- ◆ *Types V through VIII:* Care for these patients is minimal. Explain the disorder to the patient and his/her family, and help them accept the limitations imposed by the patient's particular type of glycogen storage disease.

## HYPOGLYCEMIA

Hypoglycemia is an abnormally low glucose level in the bloodstream. It occurs when glucose burns up too rapidly, when the glucose release rate falls behind tissue demands, or when excessive insulin enters the bloodstream. Hypoglycemia is classified as reactive or fasting. *Reactive*

*hypoglycemia* results from the reaction to the disposition of meals or the administration of excessive insulin. *Fasting hypoglycemia* causes discomfort during long periods of abstinence from food, for example, in the early morning before breakfast. Although hypoglycemia is a specific endocrine imbalance, its symptoms are often vague and depend on how quickly the patient's glucose levels drop. If not corrected, severe hypoglycemia may result in coma and irreversible brain damage.

### **Causes and Incidence**

Reactive hypoglycemia may take several forms. In a diabetic patient, it may result from administration of too much insulin or too much oral antidiabetic medication. In a mildly diabetic patient (or one in the early stages of diabetes mellitus), reactive hypoglycemia may result from delayed and excessive insulin production after carbohydrate ingestion. Similarly, a nondiabetic patient may develop reactive hypoglycemia from a sharp increase in insulin output after a meal. Sometimes called *postprandial hypoglycemia*, this type of reactive hypoglycemia usually disappears when the patient eats something sweet. In some patients, reactive hypoglycemia has no known cause (idiopathic reactive) or may result from gastric dumping syndrome and from impaired glucose tolerance.

Hypoglycemia is at least as common in neonates and children as it is in adults and affects 1 out of 1,000 people. Usually, infants develop hypoglycemia because of an increased number of cells per unit of body weight and because of increased demands on stored liver glycogen to support respirations, thermoregulation, and muscular activity. In full-term neonates, hypoglycemia may occur 24 to 72 hours after birth and is usually transient. In neonates who are premature or small for gestational age, the onset of hypoglycemia is much more rapid (it can occur as soon as 6 hours after birth) because of their small, immature livers, which produce much less glycogen. Maternal disorders that can produce hypoglycemia in neonates within 24 hours after birth include diabetes mellitus, toxemia, erythroblastosis, and glycogen storage disease.

### **Pathophysiology**

Fasting hypoglycemia usually results from an excess of insulin or insulin-like substance or from a decrease in counterregulatory hormones. It can be

*exogenous*, resulting from external factors such as alcohol or drug ingestion, or *endogenous*, resulting from organic problems.

Endogenous hypoglycemia may result from tumors or liver disease. Insulinomas, small islet cell tumors in the pancreas, secrete excessive amounts of insulin, which inhibit hepatic glucose production. They're generally benign (in 90% of patients). Extrapancreatic tumors, though uncommon, can also cause hypoglycemia by increasing glucose utilization and inhibiting glucose output. Such tumors occur primarily in the mesenchyma, liver, adrenal cortex, GI system, and lymphatic system. They may be benign or malignant. Among nonendocrine causes of fasting hypoglycemia are severe liver diseases, including hepatitis, cancer, cirrhosis, and liver congestion associated with heart failure. All of these conditions reduce the uptake and release of glycogen from the liver. Some endocrine causes include adrenocortical insufficiency, which contributes to hypoglycemia by reducing the production of cortisol and cortisone needed for gluconeogenesis; and pituitary insufficiency, which reduces corticotropin and growth hormone levels.

## Complication

- ◆ Permanent brain damage

## Signs and Symptoms

Signs and symptoms of reactive hypoglycemia include fatigue, malaise, nervousness, irritability, trembling, tension, headache, hunger, cold sweats, and rapid heart rate. These same clinical effects usually characterize fasting hypoglycemia. In addition, fasting hypoglycemia may also cause CNS disturbances; for example, blurry or double vision, confusion, motor weakness, hemiplegia, seizures, or coma.

In infants and children, signs and symptoms of hypoglycemia are vague. A neonate's refusal to feed may be the primary clue to underlying hypoglycemia. Associated CNS effects include tremors, twitching, weak or high-pitched cry, sweating, limpness, seizures, and coma.

## Diagnosis

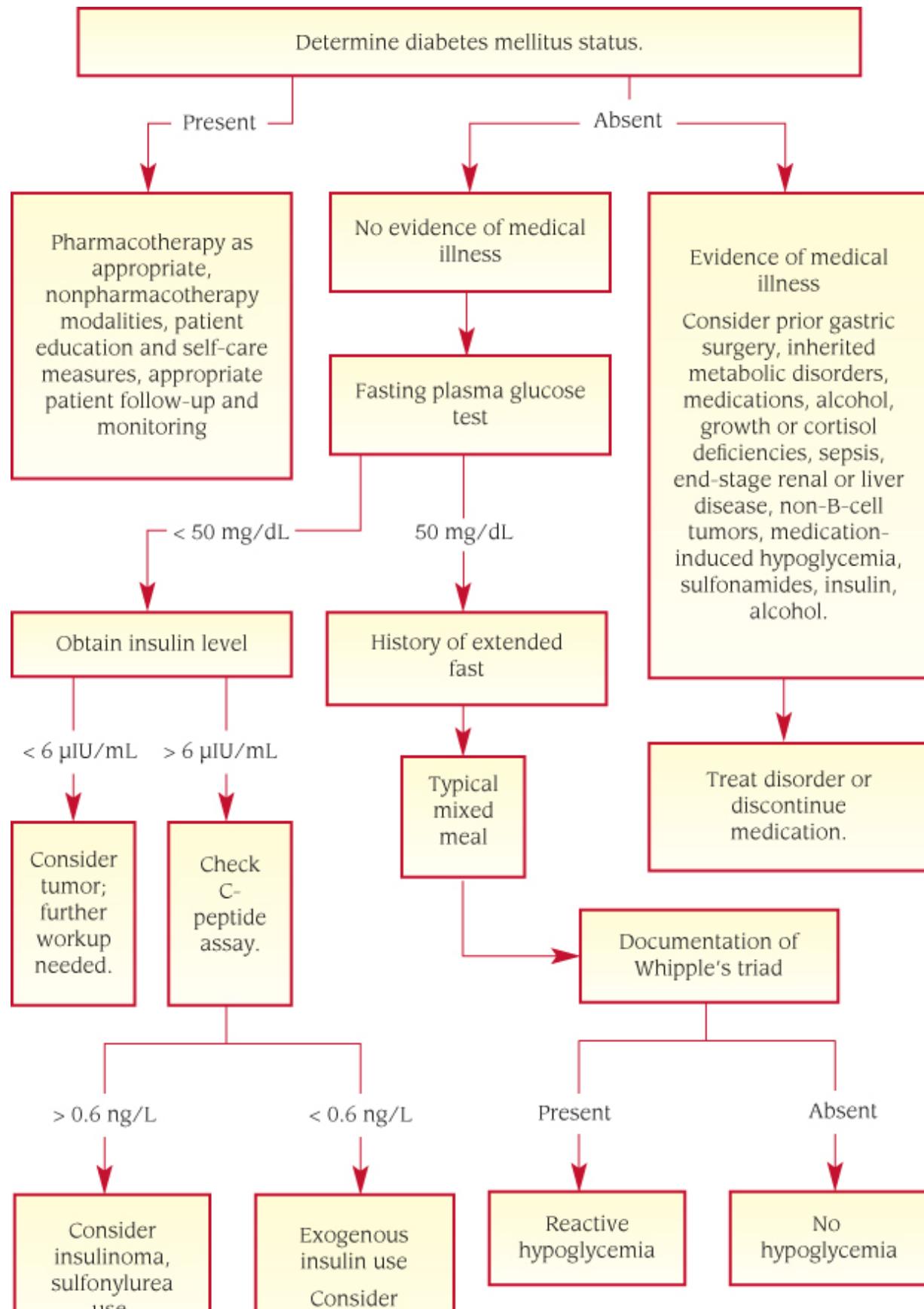
A blood glucose monitor or glucose reagent strips provide quick screening methods for determining the blood glucose level. A reading less than 50

mg/dL indicates the need for a venous blood sample. (See *Diagnosing hypoglycemia*, page 512.)



## **DIFFERENTIAL DIAGNOSIS DIAGNOSING HYPOGLYCEMIA**

THIS FLOWCHART LISTS POSSIBLE DIAGNOSTIC FINDINGS AND INTERPRETATIONS TO ASSIST WITH TREATMENT OF THE PATIENT WITH HYPOGLYCEMIA.



use.

autoimmune  
hypoglycemia,  
factitious use of  
insulin.

**D** **CONFIRMING DIAGNOSIS** *Laboratory testing confirms the diagnosis by showing decreased blood glucose levels. The following values indicate hypoglycemia:*

- ◆ *Full-term infants*
  - ◆ less than 30 mg/dL before feeding
  - ◆ less than 40 mg/dL after feeding
- ◆ *Preterm infants*
  - ◆ less than 20 mg/dL before feeding
  - ◆ less than 30 mg/dL after feeding
- ◆ *Children and adults*
  - ◆ less than 40 mg/dL before meal
  - ◆ less than 50 mg/dL after meal

In addition, a 5-hour glucose tolerance test may be administered to provoke reactive hypoglycemia. Following a 12-hour fast, laboratory testing to detect plasma insulin and plasma glucose levels may identify fasting hypoglycemia.

## Treatment

Effective treatment of reactive hypoglycemia requires dietary modification to help delay glucose absorption and gastric emptying. Usually this includes small, frequent meals; ingestion of complex carbohydrates, fiber, and fat; and avoidance of simple sugars, alcohol, and fruit drinks. The patient may also receive anticholinergic drugs to slow gastric emptying and intestinal motility and to inhibit vagal stimulation of insulin release.

For fasting hypoglycemia, surgery and drug therapy are usually required. In patients with insulinoma, tumor removal is the treatment of choice. Drug therapy may include nondiuretic thiazides such as diazoxide to inhibit

insulin secretion; streptozocin; and hormones, such as glucocorticoids and long-acting glycogen.

Therapy for neonates who have hypoglycemia or who are at risk of developing it includes preventive measures. A hypertonic solution of 10% dextrose, calculated at 5 to 10 mL/kg of body weight administered I.V. over 10 minutes and followed by 4 to 8 mg/kg/minute for maintenance, should correct a severe hypoglycemic state in neonates. To reduce the chance of hypoglycemia in high-risk neonates, they should receive feedings (either breast milk or a solution of 5% to 10% glucose and water) as soon after birth as possible.

## **Special Considerations**

- ◆ Watch for and report signs of hypoglycemia, such as poor feeding, in high-risk neonates.
- ◆ Monitor infusion of hypertonic glucose in the neonate to avoid hyperglycemia, circulatory overload, and cellular dehydration. Terminate glucose solutions gradually to prevent hypoglycemia caused by hyperinsulinemia.
- ◆ Explain the purpose and procedure for any diagnostic tests. Collect blood samples at the appropriate times, as ordered.
- ◆ Monitor the effects of drug therapy and watch for the development of any adverse effects.
- ◆ Teach the patient or family which foods to include in the diet (complex carbohydrates, fiber, fat) and which foods to avoid (simple sugars, alcohol). Refer the patient and family for dietary counseling as appropriate.

## **HEREDITARY FRUCTOSE INTOLERANCE**

Hereditary fructose intolerance is an inability to metabolize fructose. After fructose is eliminated from the diet, symptoms subside within weeks. Older children and adults with hereditary fructose intolerance have normal intelligence and apparently normal liver and kidney function.

## **Causes and Incidence**

Transmitted as an autosomal recessive trait, hereditary fructose intolerance results from a deficiency in the enzyme fructose-1-phosphate aldolase. The

enzyme operates at only 1% to 10% of its normal biological activity, thus preventing rapid uptake of fructose by the liver after ingestion of fruit or foods containing cane sugar.

In some European countries, hereditary fructose intolerance may have an incidence as high as 1 in 20,000 people.

## Pathophysiology

Affected individuals are completely asymptomatic until they ingest fructose. Thus, homozygous neonates remain clinically well until confronted with dietary sources of fructose. Although lactose is the carbohydrate base in most infant formulas, some (e.g., soy formulas) contain sucrose, a fructose-glucose disaccharide that may cause symptoms. The biochemistry of hereditary fructose intolerance is complex for two reasons: (1) Three isozymes of aldolase (A, B, C) exist, of which aldolase B is expressed exclusively in the liver, kidney, and intestine; and (2) aldolase B mediates three separate reactions (i.e., cleavage of fructose 1-phosphate [F-1-P]; cleavage of fructose 1,6-diphosphate; and condensation of the triose phosphates, glyceraldehyde phosphate, and dihydroxyacetone phosphate to form fructose 1,6-diphosphate).

In normal cellular conditions, the primary enzymatic activity of aldolase B is to cleave fructose diphosphate, which forms rather than condenses the triose phosphate compounds. Here, the enzyme is central to the glycolytic pathway. Because the reaction is reversible, aldolase B is an essential enzyme in the process of gluconeogenesis (which is, in some respects, a reversal of glycolysis). The absence of the latter function readily explains the clinical hypoglycemia in individuals with hereditary fructose intolerance.

Reduced cleavage of F-1-P leads to its cellular accumulation and fructokinase inhibition, causing free fructose accumulation in the blood. A generally accepted consequence of this sequence is a dramatic change in the ATP-adenosine monophosphate cellular ratio, with a resultant acceleration in production of uric acid. This accounts for the hyperuricemia observed during an acute episode. Competition between urate and lactate for renal tubule excretion accounts for the lactic acidemia.

The cause of severe hepatic dysfunction remains unknown but may be a manifestation of focal cytoplasmic degeneration and cellular fructose toxicity. The cause of renal tubular dysfunction also remains unclear;

patients with renal tubular dysfunction primarily present with a proximal tubular acidosis complicated by aminoaciduria, glucosuria, and phosphaturia. Thus, in an infant who is homozygous for fructose 1-aldolase deficiency, fructose ingestion triggers a cascade of biochemical events that result in severe clinical disease.

## Complications

- ◆ Kidney dysfunction
- ◆ Liver dysfunction
- ◆ Infants—failure to thrive

## Signs and Symptoms

Typically, clinical features of hereditary fructose intolerance appear shortly after dietary introduction of foods containing fructose or sucrose. Symptoms are more severe in infants than in older people and include hypoglycemia, nausea, vomiting, pallor, excessive sweating, cyanosis, and tremor. In neonates and young children, continuous ingestion of foods containing fructose may result in failure to thrive, hypoglycemia, jaundice, hyperbilirubinemia, ascites, hepatomegaly, vomiting, dehydration, hypophosphatemia, albuminuria, aminoaciduria, seizures, coma, febrile episodes, substernal pain, and anemia.

## Diagnosis

A dietary history often suggests hereditary fructose intolerance.

 **CONFIRMING DIAGNOSIS** *A fructose tolerance test (using glucose oxidase or paper chromatography to measure glucose levels) usually confirms the diagnosis. However, liver biopsy showing a deficiency in fructose-1-phosphate aldolase may be necessary for a definitive diagnosis.*

Supportive values may include decreased serum inorganic phosphorus levels. Urine studies may show fructosuria and albuminuria.

## Treatment

Treatment of hereditary fructose intolerance consists of exclusion of fructose and sucrose (cane sugar or table sugar) from the diet. Otherwise, treatment is supportive as the patient's progress is monitored.

## Special Considerations

- ◆ Tell the patient to avoid fruits containing fructose and vegetables containing sucrose (sugar beets, sweet potatoes, and peas), because sucrose is digested to glucose and fructose in the intestine. Fruits containing the least amount of fructose include strawberries, blackberries, blueberries, oranges, and grapefruits; others low in fructose are cherries, pears, bananas, grapes, and apples.
- ◆ Refer the patient and family for genetic and dietary counseling as appropriate.

## HYPERLIPOPROTEINEMIA

Hyperlipoproteinemia occurs as five distinct metabolic disorders, all of which may be inherited.

### Causes and Incidence

This disorder affects lipid transport in serum and produces varied clinical changes, from relatively mild symptoms that can be corrected by dietary management to potentially fatal pancreatitis.

### Pathophysiology

Types I and III are transmitted as autosomal recessive traits; types II, IV, and V are transmitted as autosomal dominant traits. (See *Types of hyperlipoproteinemia*, page 515.) About one in five persons with elevated plasma lipid and lipoprotein levels has hyperlipoproteinemia. It's marked by increased plasma concentrations of one or more lipoproteins. Hyperlipoproteinemia may also occur secondary to other conditions, such as diabetes, pancreatitis, hypothyroidism, or renal disease.

### Types of hyperlipoproteinemia

| Type | Causes and incidence | Diagnostic findings |
|------|----------------------|---------------------|
| I    |                      |                     |

| Type   | Causes and incidence  | Diagnostic findings  |
|--|---|--|
| (Frederickson's hyperlipoproteinemia, fat-induced hyperlipemia, idiopathic familial)               | <ul style="list-style-type: none"> <li>◆ Deficient or abnormal lipoprotein lipase, resulting in decreased or absent post-heparin lipolytic activity</li> <li>◆ Relatively rare</li> <li>◆ Present at birth</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Chylomicrons (very-low-density lipoprotein [VLDL], low-density lipoprotein [LDL], high-density lipoprotein), in plasma 14 hours or more after last meal</li> <li>◆ Highly elevated serum chylomicrons and triglyceride levels; slightly elevated serum cholesterol levels</li> <li>◆ Lower serum lipoprotein lipase levels</li> <li>◆ Leukocytosis</li> </ul> |
| <b>II</b>  |   |  |
| (familial hyperbetalipoproteinemia, essential familial hypercholesterolemia)                       | <ul style="list-style-type: none"> <li>◆ Deficient cell surface receptor that regulates LDL degradation and cholesterol synthesis, resulting in increased levels of plasma LDL over joints and pressure points</li> <li>◆ Onset between ages 10 and 30</li> </ul> | <ul style="list-style-type: none"> <li>◆ Increased plasma concentrations of LDL</li> <li>◆ Increased serum LDL and cholesterol levels</li> <li>◆ Amniocentesis shows increased LDL levels</li> </ul>   |
| <b>III</b>   |   |  |
| (familial broad-beta disease, dysbetalipoproteinemia, remnant removal disease, xanthoma tuberosum) | <ul style="list-style-type: none"> <li>◆ Unknown underlying defect results in deficient conversion of triglyceride-rich VLDL to LDL</li> <li>◆ Uncommon; usually occurs after age 20 but can occur earlier in men</li> </ul>                                      | <ul style="list-style-type: none"> <li>◆ Abnormal serum betalipoprotein</li> <li>◆ Elevated cholesterol and triglyceride levels</li> <li>◆ Slightly elevated glucose tolerance</li> <li>◆ Hyperuricemia</li> </ul>   |
| <b>IV</b>  |   |  |
| (endogenous hypertriglyceridemia, hyperbetalipoproteinemia)  | <ul style="list-style-type: none"> <li>◆ Usually occurs secondary to obesity, alcoholism, diabetes, or emotional disorders</li> <li>◆ Relatively common, especially in middle-age men</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Elevated VLDL levels</li> <li>◆ Abnormal levels of triglycerides in plasma; variable increase in serum</li> <li>◆ Normal or slightly elevated serum cholesterol levels</li> <li>◆ Mildly abnormal glucose tolerance</li> <li>◆ Family history</li> <li>◆ Early coronary artery disease</li> </ul>   |
| <b>V</b>   |   |  |

| Type   | Causes and incidence   | Diagnostic findings   |
|--|--|---|
| (mixed hypertriglyceridemia, mixed hyperlipidemia) | <ul style="list-style-type: none"> <li>◆ Defective triglyceride clearance causes pancreatitis; usually secondary to another disorder, such as obesity or nephrosis</li> <li>◆ Uncommon; onset usually occurs in late adolescence or early adulthood</li> </ul> | <ul style="list-style-type: none"> <li>◆ Chylomicrons in plasma</li> <li>◆ Elevated plasma VLDL levels</li> <li>◆ Elevated serum cholesterol and triglyceride levels</li> </ul> |

## Complications

- ◆ Coronary artery disease
- ◆ Pancreatitis

## Signs and Symptoms

- ◆ *Type I:* recurrent attacks of severe abdominal pain similar to pancreatitis, usually preceded by fat intake; abdominal spasm, rigidity, or rebound tenderness; hepatosplenomegaly, with liver or spleen tenderness; papular or eruptive xanthomas (pinkish-yellow cutaneous deposits of fat) over pressure points and extensor surfaces; lipemia retinalis (reddish-white retinal vessels); malaise; anorexia; and fever
- ◆ *Type II:* tendinous xanthomas (firm masses) on the Achilles tendons and tendons of the hands and feet, tuberous xanthomas, xanthelasma, juvenile corneal arcus (opaque ring surrounding the corneal periphery), accelerated atherosclerosis and premature coronary artery disease, and recurrent polyarthritis and tenosynovitis
- ◆ *Type III:* peripheral vascular disease manifested by claudication or tuboeruptive xanthomas (soft, inflamed, pedunculated lesions) over the elbows and knees; palmar xanthomas on the hands, particularly the fingertips; premature atherosclerosis
- ◆ *Type IV:* predisposition to atherosclerosis and early coronary artery disease, exacerbated by excessive calorie intake, obesity, diabetes, and hypertension
- ◆ *Type V:* abdominal pain (most common), pancreatitis, peripheral neuropathy, eruptive xanthomas on extensor surfaces of the arms and legs, lipemia retinalis, and hepatosplenomegaly

## Treatment

The first goal is to identify and treat any underlying problem such as diabetes. If no underlying problem exists, the primary treatment of types II, III, and IV is dietary management, especially restriction of cholesterol intake, possibly supplemented by drug therapy (cholestyramine, fenofibrate, gemfibrozil, atorvastatin, niacin) to lower plasma triglyceride or cholesterol level when diet alone is ineffective.

Type I hyperlipoproteinemia requires long-term weight reduction, with fat intake restricted to less than 20 g/day. A 20- to 40-g/day medium-chain triglyceride diet may be ordered to supplement calorie intake. The patient should also avoid alcoholic beverages, to decrease plasma triglyceride levels. The prognosis is good with treatment; without treatment, death can result from pancreatitis.

For type II, dietary management to restore normal lipid levels and decrease the risk of atherosclerosis includes restriction of cholesterol intake to less than 300 mg/day for adults and less than 150 mg/day for children; triglycerides must be restricted to less than 100 mg/day for children and adults. Diet should also be high in polyunsaturated fats. In familial hypercholesterolemia, nicotinic acid with a bile acid usually normalizes low-density lipoprotein (LDL) levels. For severely affected children, a portacaval shunt is a last resort to reduce plasma cholesterol levels. The prognosis remains poor regardless of treatment; in homozygotes, myocardial infarction usually causes death before age 30.

For type III, dietary management includes restriction of cholesterol intake to less than 300 mg/day; carbohydrates must also be restricted, while polyunsaturated fats are increased. Statins, fibrates, estrogens (in women) and niacin help lower blood lipid levels. Weight reduction is helpful. With strict adherence to prescribed diet, the prognosis is good.

For type IV, weight reduction may normalize blood lipid levels without additional treatment. Long-term dietary management includes restricted cholesterol intake, increased polyunsaturated fats, and avoidance of alcoholic beverages. Fenofibrate, gemfibrozil, and niacin may lower plasma lipid levels. The prognosis remains uncertain, however, because of predisposition to premature coronary artery disease.

The most effective treatment for type V is weight reduction and long-term maintenance of a low-fat diet. Alcoholic beverages must be avoided. Niacin, fenofibrate, gemfibrozil, and a 20- to 40-g/day medium-chain

triglyceride diet may prove helpful. The prognosis is uncertain because of the risk of pancreatitis. Increased fat intake may cause recurrent bouts of illness, possibly leading to pseudocyst formation, hemorrhage, and death.

## Special Considerations

Nursing care for hyperlipoproteinemia emphasizes careful monitoring for adverse drug effects and teaching the importance of long-term dietary management.

- ◆ Administer cholestyramine before meals or before bedtime. This drug must not be given with other medications. (See *Using bile acid sequestrants*.) Watch for adverse effects, such as nausea, vomiting, constipation, steatorrhea, rashes, and hyperchloremic acidosis. Also watch for malabsorption of other medications and fat-soluble vitamins.

### Using bile acid sequestrants

Before giving the patient a bile acid sequestrant, such as cholestyramine, to lower cholesterol levels, make certain the patient isn't taking a drug whose absorption is affected by bile acid sequestrants. For example, bile acid sequestrants decrease the absorption of diuretics such as chlorothiazide. Other drugs affected besides diuretics include:

- ◆ beta-adrenergic blockers
- ◆ digitoxin
- ◆ fat-soluble vitamins
- ◆ folic acid
- ◆ thiazides
- ◆ thyroxine
- ◆ warfarin



**ALERT** *Don't administer niacin to patients with active peptic ulcers or hepatic disease. Use with caution in patients with diabetes. In other patients, watch for adverse effects, such as flushing, pruritus, hyperpigmentation, and exacerbation of inactive peptic ulcers.*

- ◆ Urge the patient to adhere to the ordered diet (usually 1,000 to 1,500 calories/day) and to avoid excess sugar and alcoholic beverages, to minimize the intake of saturated fats (higher in meats, coconut oil), and to increase the intake of polyunsaturated fats (vegetable oils).
- ◆ Instruct the patient, for the 2 weeks preceding serum cholesterol and serum triglyceride tests, to maintain a steady weight and to adhere strictly to the prescribed diet. The patient should also fast for 12 hours preceding the test.

## **GAUCHER DISEASE**

Gaucher disease, the most common lysosomal storage disease, causes an abnormal accumulation of glucocerebrosides in reticuloendothelial cells. It occurs in three forms: type I (adult); type II (infantile); and type III (juvenile). Type II can prove fatal within 9 months of onset, usually from pulmonary involvement.

### **Causes and Incidence**

The three forms of Gaucher disease are classified by age of onset and the presence or absence of neurologic involvement. Type I, characterized by lack of neurologic involvement, is the most common form affecting both children and adults and is most prevalent in the Ashkenazi Jewish population, affecting anywhere from 1 of 500 to 1,000 births. Type II usually presents in infancy with severe neurologic involvement, resulting in seizures and CNS damage. Type II also presents with spleen and bone marrow damage. Type III typically has mild neurologic involvement and runs a slower, more favorable course. The incidence of types II and III is 1 of 50,000 to 100,000 births. The juvenile form can begin in childhood, typically in the teenage years, and causes spleen, bone marrow, and neurologic damage.

### **Pathophysiology**

Gaucher disease results from an autosomal recessive inheritance, which causes decreased activity of the enzyme glucocerebrosidase. Glucocerebrosidase deficiency leads to an accumulation of glucosylceramide in the storage compartments (lysosomes) of certain body cells. Glucosylceramide buildup occurs in the liver, spleen, bones, and bone

marrow, eventually leading to decreased production of RBCs (anemia) and thinning of the bones (osteopenia).

## Complications

- ◆ Neurologic impairment
- ◆ Portal hypertension
- ◆ Pathologic fractures
- ◆ Anemia
- ◆ Respiratory failure (type II)

## Signs and Symptoms

The key signs of all types of Gaucher disease are hepatosplenomegaly and bone lesions. In type I, bone lesions lead to thinning of cortices, pathologic fractures, collapsed hip joints and, eventually, vertebral compression. Severe episodic pain may develop in the legs, arms, and back but usually not until adolescence. (The adult form of Gaucher disease is generally diagnosed while the patient is in their teens; the word “adult” is used loosely here.) Other clinical effects of type I are fever, abdominal distention (from hypotonicity of the large bowel), respiratory problems (pneumonia or, rarely, cor pulmonale), easy bruising and bleeding, anemia and, rarely, pancytopenia. Older patients may develop a yellow pallor and brown-yellow pigmentation on the face and legs.

In type II, motor dysfunction and spasticity occur at age 6 to 7 months. Other signs of the infantile form of Gaucher disease include abdominal distention, strabismus, muscle hypertonicity, retroflexion of the head, neck rigidity, dysphagia, laryngeal stridor, hyperreflexia, seizures, respiratory distress, and easy bruising and bleeding.

Clinical effects of type III after infancy include seizures, hypertonicity, strabismus, poor coordination and mental ability and, possibly, easy bruising and bleeding.

## Diagnosis



**CONFIRMING DIAGNOSIS** Bone marrow aspiration showing Gaucher cells and direct assay of glucocerebrosidase activity, which can be performed on venous blood, confirms this diagnosis.

Supportive laboratory results include increased serum acid phosphatase level, decreased platelets and serum iron level and, in type III, abnormal EEG after infancy.

## Treatment

Treatment is mainly supportive and consists of vitamins, supplemental iron or liver extract to prevent anemia caused by iron deficiency and to alleviate other hematologic problems, blood transfusions for anemia, splenectomy for thrombocytopenia, and strong analgesics for bone pain. Injections of a replacement synthetic enzyme have proven helpful. Imiglucerase, a recombinant form of acid beta-glucuronidase has been used to treat symptomatic Gaucher disease. The FDA has recently approved velaglucerase alfa for injection to treat children and adults as a long-term enzyme replacement therapy for type I Gaucher disease. Patients receiving imiglucerase can safely be switched to velaglucerase alfa.

## Special Considerations

- ◆ In the patient confined to bed, prevent pathologic fractures by turning the patient carefully. If the patient is ambulatory, make sure that he's assisted when getting out of bed or walking.
- ◆ Observe closely for changes in pulmonary status.
- ◆ Explain all diagnostic tests and procedures to the patient or parents. Help the patient accept the limitations imposed by this disorder.
- ◆ Recommend genetic counseling for patients with a family history of Gaucher disease. Prenatal testing can determine if a fetus has the syndrome.

## PORPHYRIAS

Classification of porphyrias depends on the site of excessive porphyrin production; they may be erythropoietic (erythroid cells in bone marrow), hepatic (in the liver), or erythrohepatic (in bone marrow and liver). (See *Types of porphyria*, pages 518 and 519.) An acute episode of intermittent hepatic porphyria may cause fatal respiratory paralysis. In the other forms of porphyrias, the prognosis is good with proper treatment.

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### Types of porphyria

| <b>Porphyria</b>                | <b>Signs and symptoms</b>  | <b>Treatment</b>  |
|---------------------------------|--|---|
| <b>Erythropoietic porphyria</b> |  |   |
| <b>Günther disease</b>          | <ul style="list-style-type: none"> <li>◆ Red urine (earliest, most characteristic sign); severe cutaneous photosensitivity, leading to vesicular or bullous eruptions on exposed areas and, eventually, scarring and ulceration</li> <li>◆ Hypertrichosis</li> <li>◆ Brown-stained or red-stained teeth</li> <li>◆ Splenomegaly, hemolytic anemia</li> </ul> | <ul style="list-style-type: none"> <li>◆ Oral beta-carotene to prevent photosensitivity reactions</li> <li>◆ Anti-inflammatory ointments</li> <li>◆ Oral activated charcoal to absorb excess porphyrins</li> <li>◆ Packed red cells to inhibit erythropoiesis and excreted porphyrins</li> <li>◆ Heme therapy for recurrent attacks</li> <li>◆ Splenectomy for hemolytic anemia</li> <li>◆ Topical dihydroxyacetone and lawsone sunscreen filter</li> </ul> |
| <b>Erythrohepatic porphyria</b> |  |   |
| <b>Protoporphyria</b>           | <ul style="list-style-type: none"> <li>◆ Usually affects children</li> <li>◆ Occurs most often in males</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Photosensitive dermatitis</li> <li>◆ Hemolytic anemia</li> <li>◆ Chronic hepatic disease</li> </ul>  |
| <b>Toxic-acquired porphyria</b> | <ul style="list-style-type: none"> <li>◆ Acute colicky pain</li> <li>◆ Anorexia, nausea, vomiting</li> <li>◆ Neuromuscular weakness</li> <li>◆ Behavioral changes</li> <li>◆ Seizures, coma</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Avoidance of causative factors</li> <li>◆ Beta-carotene to reduce photosensitivity</li> </ul>  |
| <b>Hepatic porphyria</b>        |  | <ul style="list-style-type: none"> <li>◆ Chlorpromazine I.V. to relieve pain and GI symptoms</li> <li>◆ Avoidance of lead exposure</li> </ul>   |

| <b>Porphyria</b>                            | <b>Signs and symptoms</b>   | <b>Treatment</b>   |
|---|---|--|
| <b>Acute<br/>intermittent<br/>porphyria</b> | <ul style="list-style-type: none"> <li>◆ Colicky abdominal pain with fever, general malaise, and hypertension</li> <li>◆ Peripheral neuritis, behavioral changes, possibly leading to frank psychosis</li> <li>◆ Possible respiratory paralysis</li> </ul> <ul style="list-style-type: none"> <li>◆ Most common form</li> <li>◆ Affects females most often, usually between ages 15 and 40</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Chlorpromazine I.V. to relieve abdominal pain and control psychic abnormalities</li> <li>◆ Avoidance of barbiturates, sulfonamides, infections, alcohol, and fasting</li> <li>◆ Heme therapy for recurrent attacks</li> <li>◆ High-carbohydrate diet</li> </ul> |
| <b>Variegate<br/>porphyria</b>              | <ul style="list-style-type: none"> <li>◆ Skin lesions, extremely fragile skin in exposed areas</li> <li>◆ Hypertrichosis</li> <li>◆ Hyperpigmentation</li> <li>◆ Abdominal pain during acute attack</li> <li>◆ Neuropsychiatric manifestations</li> </ul> <ul style="list-style-type: none"> <li>◆ Usual onset between ages 30 and 50</li> <li>◆ Occurs almost exclusively among South African whites</li> <li>◆ Affects males and females equally</li> </ul> | <ul style="list-style-type: none"> <li>◆ Avoidance of sunlight, or wearing protective clothing when avoidance isn't possible</li> <li>◆ Heme therapy for recurrent attacks</li> </ul>  |
| <b>Porphyria<br/>cutanea tarda</b>          | <ul style="list-style-type: none"> <li>◆ Facial pigmentation</li> <li>◆ Red-brown urine</li> <li>◆ Photosensitive dermatitis</li> <li>◆ Hypertrichosis</li> </ul> <ul style="list-style-type: none"> <li>◆ Most frequent in men ages 40 to 60</li> <li>◆ Highest incidence in South Africans</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Avoidance of precipitating factors, such as alcohol and estrogens</li> <li>◆ Phlebotomy at 2-week intervals to lower serum iron level</li> </ul>  |
| <b>Hepatic porphyria</b>                    |   |  |

| <b>Porphyria</b>                 | <b>Signs and symptoms</b>  | <b>Treatment</b>  |
|----------------------------------|--|---|
| <b>Hereditary coproporphyria</b> | <ul style="list-style-type: none"> <li>◆ Asymptomatic or mild neurologic, abdominal, or psychiatric symptoms</li> <li>◆ Rare</li> <li>◆ Affects males and females equally</li> </ul> | <ul style="list-style-type: none"> <li>◆ High-carbohydrate diet</li> <li>◆ Avoidance of barbiturates</li> <li>◆ Heme therapy for recurrent attacks</li> </ul> |

## Causes and Incidence

Porphyrias are inherited as autosomal dominant traits, except for Günther disease (autosomal recessive trait) and toxic-acquired porphyria (usually from ingestion of or exposure to lead). Menstruation often precipitates acute porphyria in premenopausal women. (See *Preventing porphyria*.)



## PREVENTION PREVENTING PORPHYRIA

Precipitating factors may lead to signs and symptoms of porphyria. Encourage the patient to avoid:

- ◆ crash dieting
- ◆ fasting
- ◆ specific drugs, including alcohol, barbiturates, and estrogens
- ◆ stress
- ◆ infection

## Helpful Hints

- ◆ Stress management techniques may help because emotional stress may also precipitate an attack.
- ◆ Reduce infection risk with proper handwashing and avoiding people with known infection.
- ◆ Genetic counseling may also be of benefit to prospective parents with a family history of porphyria.

## Pathophysiology

Porphyrias are inborn errors of metabolism that affect the biosynthesis of heme (a component of hemoglobin) and cause excessive production and excretion of porphyrins or their precursors. Porphyrins, which are present in all protoplasm, figure prominently in energy storage and utilization.

## Complications

- ◆ Neurologic and hepatic dysfunction (hepatic)
- ◆ Cholelithiasis
- ◆ Coma
- ◆ Flaccid paralysis, respiratory paralysis, and death (acute intermittent)
- ◆ Hemolytic anemia (erythropoietic)

## Signs and Symptoms

Porphyrias are generally marked by photosensitivity, acute abdominal pain, and neuropathy. Hepatic porphyrias may produce a complex syndrome marked by distinct neurologic and hepatic dysfunction:

- ◆ Neurologic symptoms include chronic brain syndrome, peripheral neuropathy and autonomic effects, tachycardia, labile hypertension, severe colicky lower abdominal pain, and constipation.
- ◆ During an acute attack, fever, leukocytosis, and fluid and electrolyte imbalance may occur.
- ◆ Structural hepatic effects include fatty infiltration of the liver, hepatic sclerosis, and focal hepatocellular necrosis.
- ◆ Skin lesions may cause itching and burning, erythema, and altered pigmentation and edema in areas exposed to light. Some chronic skin changes include milia (white papules on the hands' dorsal aspects) and hirsutism on the upper cheeks and periorbital areas.

## Diagnosis

 **CONFIRMING DIAGNOSIS** Generally, diagnosis requires screening tests for porphyrins or their precursors (such as aminolevulinic acid [ALA] and porphobilinogen [PBG]) in urine, stool, blood or, occasionally, skin biopsy. A urinary lead level of 0.2 mg/L confirms toxic-acquired porphyria.

Other laboratory values may include increased serum iron levels in porphyria cutanea tarda; leukocytosis, syndrome of inappropriate

antidiuretic hormone (SIADH), and elevated bilirubin and alkaline phosphatase levels in acute intermittent porphyria.

## Treatment

Treatment for porphyrias includes avoiding overexposure to the sun and using beta-carotene to reduce photosensitivity, as well as support for acute and long-term management. Heme therapy (panhematin) is given to control recurrent attacks of acute intermittent porphyria, Günther disease, variegate porphyria, and hereditary coproporphyria. A high-carbohydrate diet decreases urinary excretion of ALA and PBG, with restricted fluid intake to inhibit release of antidiuretic hormone (ADH).

## Special Considerations

- ◆ Warn the patient to avoid excessive sun exposure, use a sunscreen when outdoors, and take a beta-carotene supplement to reduce photosensitivity.
- ◆ Encourage a high-carbohydrate diet.
- ◆ Administer beta-carotene and hemin, as ordered.
- ◆ Advise the patient to avoid drugs that may precipitate signs and symptoms of porphyrias. (See *Drugs that aggravate porphyria*.)

## Drugs that Aggravate Porphyria

Make sure the patient with porphyria doesn't receive any of the following drugs, which are known to precipitate signs and symptoms of porphyria:

- ◆ Alcohol
- ◆ Barbiturates
- ◆ Carbamazepine
- ◆ Carisoprodol
- ◆ Chloramphenicol
- ◆ Chlordiazepoxide
- ◆ Diazepam
- ◆ Ergot alkaloids
- ◆ Estrogens
- ◆ Griseofulvin
- ◆ Imipramine

- ◆ Meprobamate
- ◆ Methylsuximide
- ◆ Methyldopa
- ◆ Pentazocine
- ◆ Phenytoin
- ◆ Progesterones
- ◆ Sulfonamides
- ◆ Tolbutamide

## METABOLIC SYNDROME

Metabolic syndrome—also called *syndrome X*, *insulin resistance syndrome*, *dysmetabolic syndrome*, and *obesity syndrome*—is a cluster of conditions characterized by abdominal obesity, high blood glucose (type 2 diabetes mellitus), insulin resistance, high blood cholesterol and triglycerides, low levels of high-density lipoprotein (HDL); and high blood pressure. More than 22% of people in the United States meet three or more of these criteria, raising their risk of heart disease and stroke and placing them at high risk for dying of myocardial infarction.

### Causes and Incidence

Abdominal obesity is a strong predictor of metabolic syndrome because abdominal fat tends to be more resistant to insulin than fat in other areas. This increases the release of free fatty acids into the portal system, leading to increased apolipoprotein B, increased LDL, decreased HDL, and increased triglyceride levels. As a result, the risk of cardiovascular disease is increased.

Type 2 diabetes mellitus is a risk factor because a hallmark for metabolic syndrome is a fasting glucose level greater than 100 mg/dL. People with diabetes develop atherosclerotic heart disease at a younger age than other people. They're also at increased risk of macrovascular disease (ischemic heart disease, stroke, and peripheral vascular disease). Diabetes is a coronary artery disease risk equivalent.

Insulin resistance and dyslipidemia are also risk factors because insulin resistance leads to hyperinsulinemia, hyperglycemia, abnormal glucose and lipid metabolism, damaged endothelium, and cardiovascular disease. Insulin is also responsible for reducing the amount of free fatty acids in the

liver. However, people with insulin resistance have an increased amount of free fatty acids reaching the liver, resulting in high triglycerides and LDLs and producing an abnormal endothelium and atherosclerosis.

High blood pressure is a risk factor because the combination of insulin resistance, hyperinsulinemia, and abdominal obesity leads to hypertension and its harmful cardiovascular effects. Moreover, insulin resistance promotes salt sensitivity in people with high blood pressure.

Women who have a history of polycystic ovarian syndrome are also at increased risk for developing metabolic syndrome.

Research also indicates that there may be a genetic predisposition to metabolic syndrome.

## **Pathophysiology**

In the normal digestion process, the intestines break down food into its basic components, one of which is glucose. Glucose provides energy for cellular activity, while excess glucose is stored in cells for future use. Insulin, a hormone secreted in the pancreas, guides glucose into storage cells. However, in people with metabolic syndrome, glucose is insulin-resistant and doesn't respond to insulin's attempt to guide it into storage cells. Excess insulin is then required to overcome this resistance. This excess in quantity and force of insulin causes damage to the lining of the arteries, promotes fat storage deposits, and prevents fat breakdown. This series of events can lead to diabetes, blood clots, and coronary events.

## **Complications**

- ◆ Coronary artery disease
- ◆ Diabetes mellitus
- ◆ Hyperlipidemia

## **Signs and Symptoms**

Assessment commonly reveals a history of hypertension, abdominal obesity, sedentary lifestyle, poor diet, and a family history of metabolic syndrome. Physical findings include abdominal obesity (evidenced by a waist of more than 40" [101.6 cm] in men and 35" [88.9 cm] in women), blood pressure 130/85 mm Hg or higher, and a fasting blood glucose level that's 100 mg/dL or higher. The patient may feel tired, especially after eating, and may have difficulty losing weight. If left untreated, such

complications as coronary artery disease, diabetes, hyperlipidemia, and premature death may develop.

## Diagnosis

Blood studies commonly indicate elevated blood glucose levels, hyperinsulinemia, and elevated serum uric acid. Lipid profile studies reveal elevated LDL levels, low HDL levels, and elevated triglycerides. Further diagnostic procedures are nonspecific, but may be performed to detect hypertension, diabetes, hyperlipidemia, and hyperinsulinemia.

## Treatment

Lifestyle modification, focusing on weight reduction and exercise, is an important part of the treatment regimen. Modest weight reduction through diet and exercise considerably improves hemoglobin A<sub>1c</sub> levels, reduces insulin resistance, improves blood lipid levels, and decreases blood pressure—all elements of metabolic syndrome. Recent studies have shown that in patients with impaired glucose tolerance, losing an average of 7% of body weight reduced the risk of developing type 2 diabetes by 58%. A long-term target goal for BMI should aim for less than 25 kg/m<sup>2</sup>.

To improve cardiovascular health, a diet rich in vegetables, fruits, whole grains, fish, and low-fat dairy products combined with regular exercise is recommended. Moreover, nutrient-dense, low-energy foods should replace low-nutrient, high-calorie foods. A healthy diet should also be low in saturated fat, trans fat, cholesterol, and sodium. (See *Therapeutic lifestyle-change diet*, page 522.)

### Therapeutic lifestyle-change diet

The therapeutic lifestyle-change diet is low in saturated fats and cholesterol to reduce blood cholesterol levels and prevent development of heart disease and its complications.

| <b>Nutrient</b>     | <b>Recommended intake</b>   |
|---------------------|-----------------------------|
| Saturated fat*      | <7% of total calories       |
| Polyunsaturated fat | Up to 10% of total calories |

| <b>Nutrient</b>          | <b>Recommended intake</b>   |
|--------------------------|---|
| Monounsaturated fat      | Up to 20% of total calories   |
| Total fat                | 25% to 35% of total calories  |
| Carbohydrate**           | 50% to 60% of total calories  |
| Fiber                    | 20 to 30 g/day  |
| Protein                  | ~15% of total calories  |
| Cholesterol < 200 mg/day | Sodium ≤2,400 mg/day  |
| Total calories***        | Balance energy intake and expenditure to maintain desirable body weight and prevent weight gain |

\* Trans fatty acids are another low-density lipoprotein-raising fat that should be minimized or avoided.

\*\* Carbohydrates should be derived predominantly from foods rich in complex carbohydrates, including grains—especially whole grains, fruits, and vegetables.

\*\*\* Daily expenditure should include at least moderate physical activity (contributing about 200 kcal/day).

Source: From National Heart, Lung, and Blood Institute; National Institutes of Health; U.S. Department of Health and Human Services.

A regular exercise program of moderate physical activity, in addition to dietary modifications, promotes weight loss, improves insulin sensitivity, and reduces blood glucose levels. According to the Surgeon General's Report on Physical Activity and Health, a person should exercise moderately for a minimum of 30 minutes on most (if not all) days of the week. The selected exercise program should improve cardiovascular conditioning, increase strength through resistance training, and improve flexibility.

Medications may be used in the treatment of metabolic syndrome for patients who have a BMI of  $27 \text{ kg/m}^2$  or greater in the presence of other risk factors (such as diabetes, hypertension, and hyperlipidemia) or for patients with a BMI of  $30 \text{ kg/m}^2$  or greater without other risk factors. Weight-loss drugs may also be added to lifestyle changes if the patient hasn't achieved significant weight loss after 12 weeks.

Pharmacologic treatment may also be indicated. Phentermine is used for short-term treatment of obesity in conjunction with diet and exercise.

Surgical treatment of obesity, such as through gastric bypass procedures, produces a greater degree and duration of weight loss than other therapies and improves or resolves most of the factors of metabolic syndrome.

Candidates for surgical intervention include patients with a BMI greater than 40 kg/m<sup>2</sup> or those with a BMI greater than 35 kg/m<sup>2</sup> with obesity-related medical conditions. Gastric bypass procedures produce permanent weight loss in the majority of patients.

## **Special Considerations**

- ◆ Monitor the patient's blood pressure, blood glucose, blood cholesterol, and insulin levels.
- ◆ Because research indicates that longer lifestyle modification programs are associated with improved weight-loss maintenance, encourage patients with metabolic syndrome to begin an exercise and weight-loss program with a friend or family member. Assist the patient in exploring options and support their efforts.
- ◆ To improve compliance, schedule frequent follow-up appointments with the patient. At that time, review food diaries and exercise logs. Be positive and promote active participation and partnership in the treatment plan.
- ◆ A patient planning gastric bypass surgery should receive psychological and nutritional counseling before and after the surgery to assist with diet and lifestyle changes.

## **Homeostatic Imbalance**

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### **POTASSIUM IMBALANCE**

Potassium, a cation that's the dominant cellular electrolyte, facilitates contraction of both skeletal and smooth muscles—including myocardial contraction—and figures prominently in nerve impulse conduction, acid–base balance, enzyme action, and cell membrane function. Because serum potassium level has such a narrow range (3.5 to 5 mEq/L), a slight deviation in either direction can produce profound clinical consequences.

### **Causes and Incidence**

Because many foods contain potassium, hypokalemia seldom results from a dietary deficiency. Instead, potassium loss may result from:

- ◆ excessive GI losses, such as diarrhea, dehydration, anorexia, or chronic laxative abuse (Vomiting and gastric suction cause dehydration, resulting in hyperaldosteronism [sodium retention and potassium excretion occur].)
- ◆ trauma (injury, burns, or surgery), in which damaged cells release potassium, which enters serum or ECF, to be excreted in the urine
- ◆ chronic renal disease, with tubular potassium wasting
- ◆ certain drugs, especially potassium-wasting diuretics, steroids, and certain sodium-containing antibiotics (carbenicillin)
- ◆ acid-base imbalances, which cause potassium shifting into cells without true depletion in alkalosis
- ◆ prolonged potassium-free I.V. therapy
- ◆ hyperglycemia, causing osmotic diuresis and glycosuria
- ◆ Cushing syndrome, primary hyperaldosteronism, excessive licorice ingestion, and severe serum magnesium deficiency
- ◆ refeeding syndrome

Hyperkalemia results from the kidneys' inability to excrete excessive amounts of potassium infused I.V. or administered orally; from decreased urine output, renal dysfunction, or renal failure; or the use of potassium-sparing diuretics, such as triamterene, by patients with renal disease. It may also result from any injuries or conditions that release cellular potassium or favor its retention, such as burns, crushing injuries, failing renal function, adrenal gland insufficiency, dehydration, or diabetic acidosis.

## **Pathophysiology**

Paradoxically, both hypokalemia (potassium deficiency) and hyperkalemia (potassium excess) can lead to muscle weakness and flaccid paralysis, because both create an ionic imbalance in neuromuscular tissue excitability. Both conditions also diminish excitability and conduction rate of the heart muscle, which may lead to cardiac arrest. (See *Clinical effects of potassium imbalance*.)

## **Clinical Effects of Potassium Imbalance**

**Dysfunction    Hypokalemia**

**Hyperkalemia**

| <b>Dysfunction</b> | <b>Hypokalemia</b>  | <b>Hyperkalemia</b>  |
|--------------------|---|--|
| Acid–base balance  | ◆ Metabolic alkalosis   | ◆ Metabolic acidosis   |
| Cardiovascular     | ◆ Dizziness, hypotension, arrhythmias, electrocardiogram (ECG) changes (flattened T waves, elevated U waves, depressed ST segment), cardiac arrest (with serum potassium levels <2.5 mEq/L) | ◆ Tachycardia and later bradycardia, ECG changes (tent and elevated T waves, widened QRS complex, prolonged PR interval, flattened or absent P waves, depressed ST segment), cardiac arrest (with levels >7 mEq/L) |
| Gastrointestinal   | ◆ Nausea, vomiting, anorexia, diarrhea, abdominal distention, paralytic ileus, or decreased peristalsis   | ◆ Nausea, diarrhea, abdominal cramps   |
| Genitourinary      | ◆ Polyuria  | ◆ Oliguria, anuria   |
| Musculoskeletal    | ◆ Muscle weakness and fatigue, leg cramps   | ◆ Muscle weakness, flaccid paralysis   |
| Neurologic         | ◆ Malaise, irritability, confusion, mental depression, speech changes, decreased reflexes, respiratory paralysis  | ◆ Hyperreflexia progressing to weakness, numbness, tingling, flaccid paralysis   |

## Complications

- ◆ Muscle weakness
- ◆ Flaccid paralysis
- ◆ Cardiac arrest

## Signs and Symptoms

Usually symptoms of low potassium are mild, sometimes vague. These include weakness, numbness/tingling, nausea/vomiting, abdominal cramps, hypotension, palpitations, and changes in behavior.

Patients with high potassium may be asymptomatic or have mild symptoms. They may report vague symptoms, such as nausea, tingling, muscle weakness, and fatigue. Symptoms may be more significant, such as slow and weak pulse, and cardiac standstill. Patients may not recognize symptoms until potassium is very high (>7.0 mEq/L). A slow rise is often better tolerated than a rapid rise.

## Diagnosis



**CONFIRMING DIAGNOSIS** Serum potassium levels less than 3.5 mEq/L confirm hypokalemia; serum levels greater than 5 mEq/L confirm hyperkalemia.

Additional tests may be necessary to determine the imbalance's underlying cause. Hypokalemia is also associated with hypomagnesemia, so further study of other electrolytes is warranted.

## Treatment

For hypokalemia, replacement therapy with potassium chloride (I.V. or orally) is the primary treatment. When diuresis is necessary, spironolactone, a potassium-sparing diuretic, may be administered concurrently with a potassium-wasting diuretic to minimize potassium loss. Hypokalemia can be prevented by giving a maintenance dose of potassium I.V. to patients who may not take anything by mouth and to others predisposed to potassium loss.

For hyperkalemia, rapid infusion of 10% calcium gluconate decreases myocardial irritability and temporarily prevents cardiac arrest but doesn't correct serum potassium excess; it's also contraindicated in patients receiving cardiac glycosides. As an emergency measure, sodium bicarbonate I.V. increases pH and causes potassium to shift back into the cells. Insulin and 10% to 50% glucose I.V. also move potassium back into cells. Infusions should be followed by dextrose 5% in water because infusion of 10% to 15% glucose will stimulate endogenous insulin secretion. Sodium polystyrene sulfonate with 70% sorbitol produces exchange of sodium ions for potassium ions in the intestine. Hemodialysis or peritoneal dialysis also aids in removal of excess potassium.

## Special Considerations

For hypokalemia:

- ◆ Check serum potassium and other electrolyte levels in patients apt to develop potassium imbalance and in those requiring potassium replacement; they risk overcorrection to hyperkalemia.
- ◆ Assess intake and output carefully. Remember, the kidneys excrete 80% to 90% of ingested potassium. Never give supplementary potassium to a patient whose urine output is below 600 mL/day. Also, measure GI loss from suctioning or vomiting.

- ◆ Administer slow-release potassium or dilute oral potassium supplements in 4 oz (118 mL) or more of water or other fluid to reduce gastric and small-bowel irritation. Determine the patient's chloride level. As ordered, give a potassium chloride supplement if the level is low and potassium gluconate if it's normal.
- ◆ Give potassium I.V. only after it's diluted in solution (usually, 10 mEq/100 mL of fluid); potassium is very irritating to vascular, subcutaneous, and fatty tissues and may cause phlebitis or tissue necrosis if it infiltrates. Infuse slowly (no more than 20 mEq/L/hour through central administration or 10 mEq/hour through peripheral administration) to prevent hyperkalemia.



**ALERT** *Never administer by I.V. push or bolus; it may cause cardiac arrest.*

- ◆ Carefully monitor patients receiving cardiac glycosides because hypokalemia enhances the action of these drugs and may produce signs of digoxin toxicity (anorexia, nausea, vomiting, blurred vision, and arrhythmias).
- ◆ To prevent hypokalemia, instruct patients (especially those predisposed to hypokalemia due to long-term diuretic therapy) to include in their diet foods rich in potassium—oranges, bananas, tomatoes, milk, dried fruits, apricots, peanuts, and dark green, leafy vegetables.



**ALERT** *Monitor the patient's cardiac rhythm and respond to any irregularities immediately.*

For hyperkalemia:

- ◆ As in hypokalemia, frequently monitor serum potassium and other electrolyte levels, and carefully record intake and output.
- ◆ Administer sodium polystyrene sulfonate orally or rectally (by retention enema) in patients with significant potassium elevations because of intravascular sodium shifting. Watch for signs of hypokalemia with prolonged use and for clinical effects of hypoglycemia (muscle weakness, syncope, hunger, and diaphoresis) with repeated insulin and glucose treatment.

- ◆ Watch for signs of hyperkalemia in predisposed patients, especially those with poor urine output or those receiving potassium supplements orally or I.V. Administer no more than 10 to 20 mEq/L of potassium chloride per hour; check the I.V. infusion site for signs of phlebitis or infiltration of potassium into tissues. Also, before giving a blood transfusion, check to see how long ago the blood was donated; cell hemolysis in older blood releases potassium. Infuse only *fresh* blood for patients with average to high serum potassium levels.
- ◆ Watch for and report cardiac arrhythmias.

## SODIUM IMBALANCE

Although the body requires only 2 to 4 g of sodium daily, most Americans consume 6 to 10 g daily (mostly sodium chloride, as table salt), excreting excess sodium through the kidneys and skin.

A low-sodium diet or excessive use of diuretics may induce hyponatremia (decreased serum sodium concentration); dehydration may induce hypernatremia (increased serum sodium concentration).

### Causes and Incidence

Hyponatremia can result from:

- ◆ excessive GI loss of water and electrolytes due to vomiting, suctioning, or diarrhea; excessive perspiration or fever; use of potent diuretics; or tap-water enemas (When such losses decrease circulating fluid volume, increased secretion of ADH promotes maximum water reabsorption, which further dilutes serum sodium. These factors are especially likely to cause hyponatremia when combined with excessive intake of free water.)
- ◆ excessive drinking of water, infusion of I.V. dextrose in water without other solutes, malnutrition or starvation, or a low-sodium diet, usually in combination with one of the other causes
- ◆ trauma, surgery (wound drainage), or burns, which cause sodium to shift into damaged cells
- ◆ adrenal gland insufficiency (Addison disease) or hypoaldosteronism
- ◆ cirrhosis of the liver with ascites
- ◆ SIADH, resulting from brain tumor, stroke, pulmonary disease, or neoplasm with ectopic ADH production. Certain drugs, such as

chlorpropamide and clofibrate, may produce an SIADH-like syndrome

Causes of hypernatremia include:

- ◆ decreased water intake (When severe vomiting and diarrhea cause water loss that exceeds sodium loss, serum sodium levels rise, but overall ECF volume decreases.)
- ◆ excess adrenocortical hormones, as in Cushing syndrome
- ◆ ADH deficiency (diabetes insipidus)
- ◆ salt intoxication (less common), which may be produced by excessive ingestion of table salt

## Pathophysiology

Sodium is the major cation (90%) in ECF; potassium, the major cation in intracellular fluid. During repolarization, the sodium-potassium pump continually shifts sodium into the cells and potassium out of the cells; during depolarization, it does the reverse. Sodium cation functions include maintaining tonicity and concentration of ECF, acid-base balance (reabsorption of sodium ion and excretion of hydrogen ion), nerve conduction and neuromuscular function, glandular secretion, and water balance.

## Complications

- ◆ Seizures
- ◆ Coma
- ◆ Permanent neurologic damage

## Signs and Symptoms

Sodium imbalance has profound physiologic effects and can induce severe CNS, cardiovascular, and GI abnormalities. For example, hyponatremia may cause renal dysfunction or, if serum sodium loss is abrupt or severe, seizures; hypernatremia may produce pulmonary edema, circulatory disorders, and decreased level of consciousness. (See *Clinical effects of sodium imbalance*.)

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### Clinical effects of sodium imbalance

| <b>Dysfunction</b> | <b>Hyponatremia</b>  | <b>Hypernatremia</b>  |
|--------------------|--|---|
| Cardiovascular     | <ul style="list-style-type: none"> <li>◆ Hypotension; tachycardia; with severe deficit, vasomotor collapse, thready pulse</li> </ul> | <ul style="list-style-type: none"> <li>◆ Hypertension, tachycardia, pitting edema, excessive weight gain</li> </ul>                 |
| Cutaneous          | <ul style="list-style-type: none"> <li>◆ Cold, clammy skin; decreased skin turgor</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Flushed skin; dry, sticky mucous membranes</li> </ul>                                      |
| ◆ Gastrointestinal | <ul style="list-style-type: none"> <li>◆ Nausea, vomiting, abdominal cramps</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Rough, dry tongue; intense thirst</li> </ul>   |
| ◆ Genitourinary    | <ul style="list-style-type: none"> <li>◆ Oliguria or anuria</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Oliguria</li> </ul>  |
| ◆ Neurologic       | <ul style="list-style-type: none"> <li>◆ Anxiety, headaches, muscle twitching and weakness, confusion, seizures</li> </ul>           | <ul style="list-style-type: none"> <li>◆ Fever, agitation, restlessness, seizures</li> </ul>  |
| ◆ Respiratory      | <ul style="list-style-type: none"> <li>◆ Cyanosis with severe deficiency</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Dyspnea, respiratory arrest, and death (from dramatic rise in osmotic pressure)</li> </ul> |

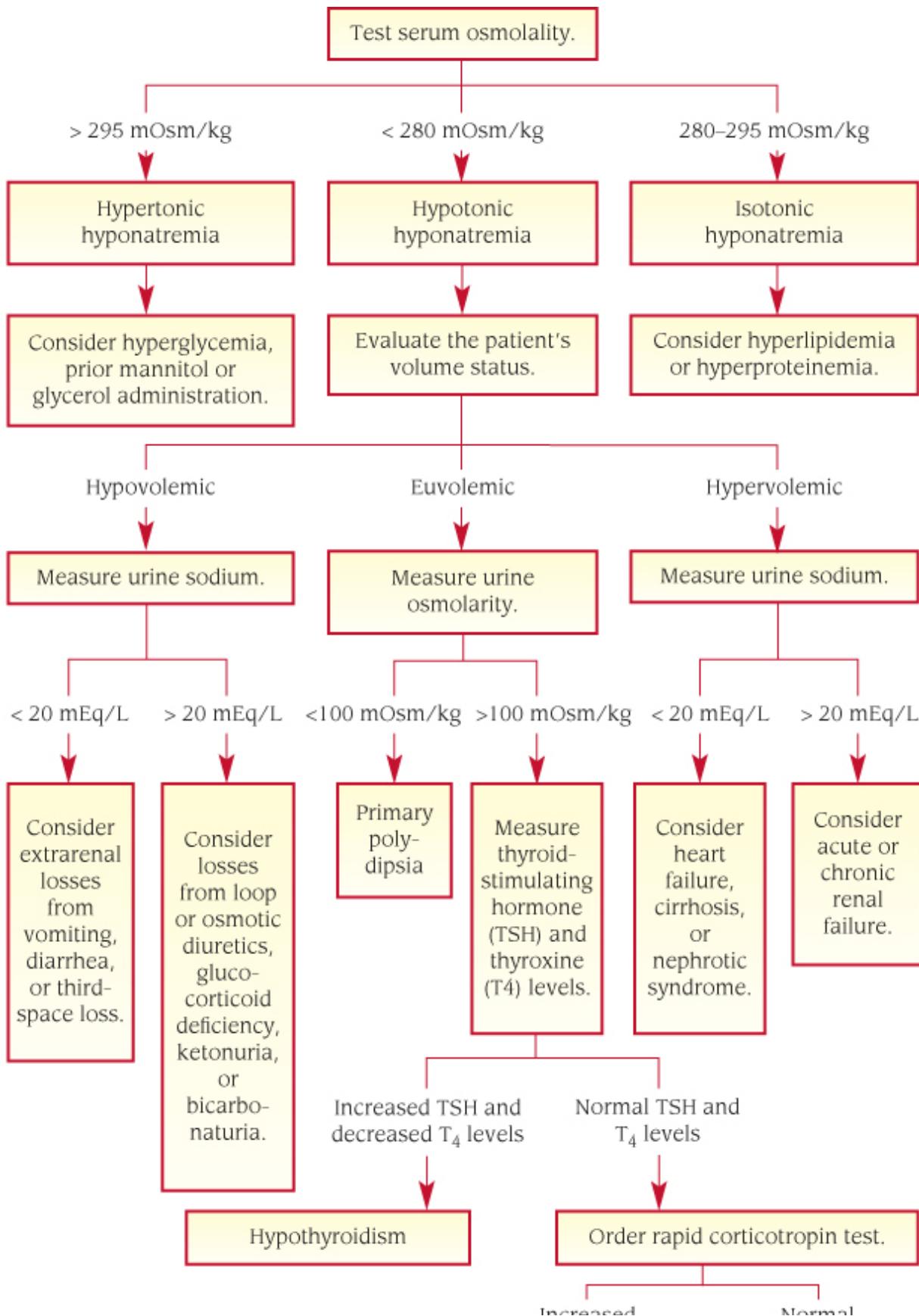
## Diagnosis

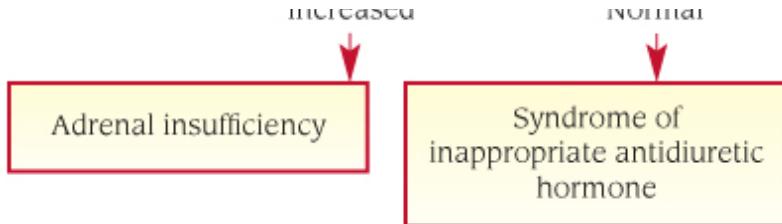
Hyponatremia is defined as a serum sodium level less than 135 mEq/L; hypernatremia, as a serum sodium level greater than 145 mEq/L. However, additional laboratory studies are necessary to determine etiology and to differentiate between a true deficit and an apparent deficit due to sodium shift or to hypervolemia or hypovolemia. In true hyponatremia, supportive values include urine sodium greater than 100 mEq/24 hours, with low serum osmolality; in true hypernatremia, urine sodium level is less than 40 mEq/24 hours, with high serum osmolality. (See *Diagnosing hyponatremia*, page 526.)



## DIFFERENTIAL DIAGNOSIS DIAGNOSING HYPONATREMIA

THIS FLOWCHART LISTS POSSIBLE DIAGNOSTIC FINDINGS AND INTERPRETATIONS TO ASSIST IN TREATING A PATIENT WITH HYPONATREMIA.





## Treatment

Therapy for mild hyponatremia usually consists of restricted free water intake when it's due to hemodilution, SIADH, or such conditions as heart failure, cirrhosis of the liver, and renal failure. If fluid restriction alone fails to normalize serum sodium levels, demeclocycline or lithium, which blocks ADH action in the renal tubules, can be used to promote water excretion. In extremely rare instances of severe symptomatic hyponatremia, when serum sodium levels fall below 110 mEq/L, treatment may include infusion of 3% or 5% saline solution.

Treatment with saline infusion requires careful monitoring of venous pressure to prevent potentially fatal circulatory overload. The aim of treatment of secondary hyponatremia is to correct the underlying disorder.

Primary treatment of hypernatremia is administration of salt-free solutions (such as dextrose in water) to return serum sodium levels to normal, followed by infusion of half-normal saline solution to prevent hyponatremia. Other measures include a sodium-restricted diet and discontinuation of drugs that promote sodium retention.

## Special Considerations

When managing the patient with hyponatremia:

- ◆ Watch for and report extremely low serum sodium and accompanying serum chloride levels. Monitor urine specific gravity and other laboratory results. Record fluid intake and output accurately, and weigh the patient daily.
- ◆ During administration of iso-osmolar or hyperosmolar saline solution, watch closely for signs of hypervolemia (dyspnea, crackles, engorged jugular or hand veins). Report conditions that may cause excessive sodium loss (diaphoresis or prolonged diarrhea or vomiting, and severe burns).

- ◆ Refer the patient on maintenance dosage of diuretics to a dietitian for instruction about dietary sodium intake.



**PREVENTION** *To prevent hyponatremia, administer iso-osmolar solutions.*

When managing the patient with hypernatremia:

- ◆ Measure serum sodium levels at least every 6 hours until stabilized. Monitor vital signs for changes, especially for rising pulse rate. Watch for signs of hypervolemia, especially in the patient receiving I.V. fluids.
- ◆ Record fluid intake and output accurately, checking for body fluid loss. Weigh the patient daily.
- ◆ Obtain a drug history to check for drugs that promote sodium retention.
- ◆ Explain the importance of sodium restriction and teach the patient how to plan a low-sodium diet. Closely monitor the serum sodium levels of high-risk patients.

## CALCIUM IMBALANCE

Calcium plays an indispensable role in cell permeability, bone and teeth formation, blood coagulation, transmission of nerve impulses, and normal muscle contraction. Nearly all (99%) of the body's calcium is found in the bones. The remaining 1% exists in the blood, with 50% of the remainder bound to plasma proteins and 40% ionized or free.

### Causes and Incidence

Common causes of hypocalcemia include:

- ◆ inadequate intake of calcium and vitamin D, in which inadequate levels of vitamin D inhibit intestinal absorption of calcium
- ◆ hypoparathyroidism as a result of injury, disease, or surgery that decreases or eliminates secretion of PTH, which is needed for calcium absorption and normal serum calcium levels
- ◆ malabsorption or loss of calcium from the GI tract, caused by increased intestinal motility from severe diarrhea or laxative abuse; can also result from inadequate levels of vitamin D or PTH, or a reduction in gastric acidity, decreasing the solubility of calcium salts

- ◆ severe infections or burns, in which diseased and burned tissue traps calcium from the ECF
- ◆ overcorrection of acidosis, resulting in alkalosis, which causes decreased ionized calcium and induces symptoms of hypocalcemia
- ◆ pancreatic insufficiency, which may cause malabsorption of calcium and subsequent calcium loss in feces. In pancreatitis, participation of calcium ions in saponification contributes to calcium loss
- ◆ renal failure, resulting in excessive excretion of calcium secondary to increased retention of phosphate
- ◆ hypomagnesemia, which causes decreased PTH secretion and blocks the peripheral action of that hormone

Causes of hypercalcemia include the following:

- ◆ hyperparathyroidism, which increases serum calcium levels by promoting calcium absorption from the intestine, resorption from bone, and reabsorption from the kidneys
- ◆ hypervitaminosis D, which can promote increased absorption of calcium from the intestine
- ◆ tumors, which raise serum calcium levels by destroying bone or by releasing PTH or a PTH-like substance, osteoclast-activating factor, prostaglandins and, perhaps, a vitamin D-like sterol
- ◆ multiple fractures and prolonged immobilization, which release bone calcium and raise the serum calcium level
- ◆ multiple myeloma, which promotes loss of calcium from bone

Other causes include milk-alkali syndrome, sarcoidosis, hyperthyroidism, adrenal insufficiency, thiazide diuretics, and loss of serum albumin secondary to renal disease.

## Complications

- ◆ Laryngeal spasm, tetany, seizures and, possibly, respiratory arrest (hypocalcemia)
- ◆ Coma and cardiac arrest (hypercalcemia)

## Pathophysiology

The ionized calcium in the serum is critical to healthy neurologic function. The parathyroid glands regulate ionized calcium and determine its

resorption into bone, absorption from the GI mucosa, and excretion in urine and feces. Severe calcium imbalance requires emergency treatment because a deficiency (hypocalcemia) can lead to tetany and seizures; an excess (hypercalcemia), to cardiac arrhythmias and coma. (See *Clinical effects of calcium imbalance*, page 527.)

## Clinical effects of calcium imbalance

| <b>Dysfunction</b> | <b>Hypocalcemia</b>   | <b>Hypercalcemia</b>   |
|--------------------|---|--|
| Cardiovascular     | ◆ Arrhythmias, hypotension  | ◆ Signs of heart block, cardiac arrest in systole, hypertension                  |
| Gastrointestinal   | ◆ Increased GI motility, diarrhea   | ◆ Anorexia, nausea, vomiting, constipation, dehydration, polydipsia              |
| Musculoskeletal    | ◆ Paresthesia (tingling and numbness of the fingers), tetany or painful tonic muscle spasms, facial spasms, abdominal cramps, muscle cramps, spasmodic contractions | ◆ Weakness, muscle flaccidity, bone pain, osteoporosis, pathologic fractures     |
| Neurologic         | ◆ Anxiety, irritability, twitching around mouth, laryngospasm, seizures, Chvostek sign, Trousseau sign  | ◆ Drowsiness, lethargy, headaches, depression or apathy, irritability, confusion |
| Other              | ◆ Blood-clotting abnormalities  | ◆ Renal polyuria, flank pain and, eventually, azotemia                           |

## Complications

- ◆ Laryngeal spasm, tetany, seizures and, possibly, respiratory arrest (hypocalcemia)
- ◆ Coma and cardiac arrest (hypercalcemia)

## Signs and Symptoms

Calcium deficit causes nerve fiber irritability and repetitive muscle spasms. Consequently, characteristic symptoms of hypocalcemia include perioral paresthesia, twitching, carpopedal spasm, tetany, seizures and, possibly, cardiac arrhythmias. Chvostek sign and Trousseau sign are reliable indicators of hypocalcemia. (See *Trousseau sign*. Also see *Chvostek sign*, page 529.)

## Trousseau Sign

To check for Trousseau sign, apply a blood pressure cuff to the patient's arm. A carpopedal spasm that causes thumb adduction and phalangeal extension, as shown, confirms tetany.



## Chvostek Sign

To check for Chvostek sign, tap the facial nerve above the mandibular angle, adjacent to the earlobe. A facial muscle spasm that causes the patient's upper lip to twitch, as shown, confirms tetany.



Clinical effects of hypercalcemia include muscle weakness, decreased muscle tone, lethargy, anorexia, constipation, nausea, vomiting, dehydration, polydipsia, and polyuria. Severe hypercalcemia (serum levels that exceed 15 mg/dL) may produce cardiac arrhythmias and, eventually, coma.

## Diagnosis

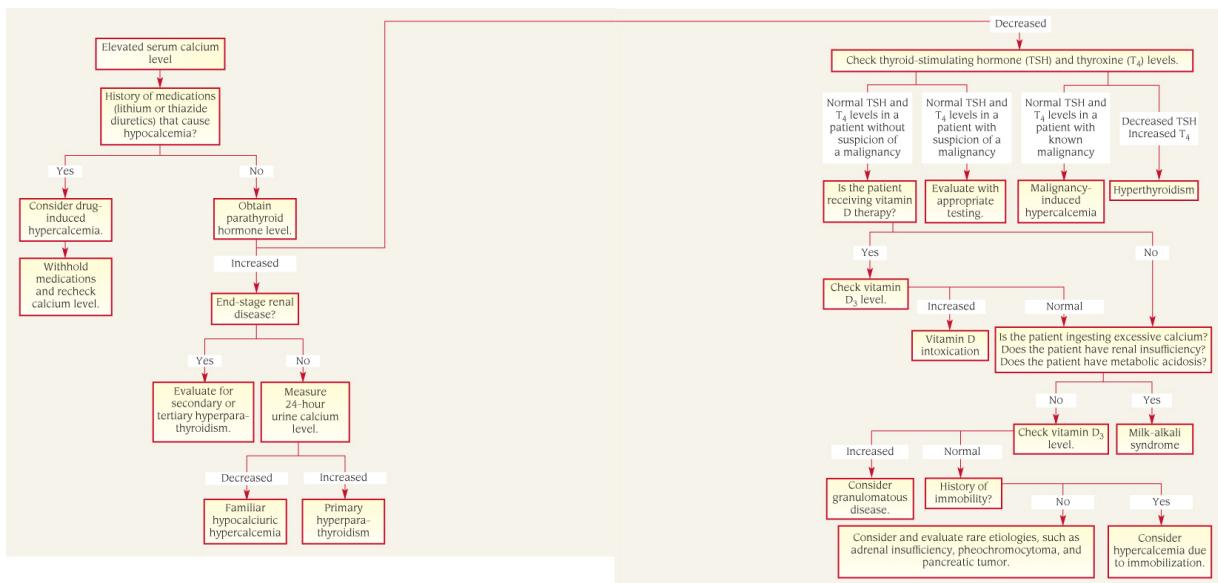
**Dx CONFIRMING DIAGNOSIS** *A serum calcium level less than 8.5 mg/dL confirms hypocalcemia; a level more than 10.5 mg/dL confirms hypercalcemia. (However, because approximately one-half of serum calcium is bound to albumin, changes in serum protein must be considered when interpreting serum calcium levels. A common conversion formula is calcium corrected = calcium actual + 0.8 × [4.0 – albumin level]. Ionized calcium levels are 4.65 to 5.28 mg/dL and are a measure of the fraction of serum calcium in ionized form.)*

The Sulkowitch urine test shows increased calcium precipitation in hypercalcemia. In hypocalcemia, an electrocardiogram (ECG) reveals lengthened QT interval, prolonged ST segment, and arrhythmias; in hypercalcemia, shortened QT interval and heart block. (See *Diagnosing hypercalcemia*, pages 530 and 531.)



## DIFFERENTIAL DIAGNOSIS DIAGNOSING HYPERCALCEMIA

THIS FLOWCHART LISTS POSSIBLE DIAGNOSTIC FINDINGS AND INTERPRETATIONS TO ASSIST WITH TREATMENT OF THE PATIENT WITH HYPERCALCEMIA.



## Treatment

Treatment varies and requires correction of the acute imbalance, followed by maintenance therapy and correction of the underlying cause. Mild hypocalcemia may require nothing more than an adjustment in diet to allow adequate intake of calcium, vitamin D, and protein, possibly with oral calcium supplements. Acute hypocalcemia is an emergency that needs immediate correction by I.V. administration of calcium gluconate or calcium chloride. Chronic hypocalcemia also requires vitamin D supplements to facilitate GI absorption of calcium. To correct mild

deficiency states, the amounts of vitamin D in most multivitamin preparations are adequate. For severe deficiency, vitamin D is used in four forms: ergocalciferol (vitamin D<sub>2</sub>), cholecalciferol (vitamin D<sub>3</sub>), calcitriol, and dihydrotachysterol, a synthetic form of vitamin D<sub>2</sub>.

Treatment of hypercalcemia primarily eliminates excess serum calcium through hydration with normal saline solution, which promotes calcium excretion in the urine. Loop diuretics, such as ethacrynic acid and furosemide, also promote calcium excretion. (Thiazide diuretics are contraindicated in hypercalcemia because they inhibit calcium excretion.) Corticosteroids, such as prednisone and hydrocortisone, are helpful in treating sarcoidosis, hypervitaminosis D, and certain tumors. Plicamycin can also lower serum calcium levels and is especially effective against hypercalcemia secondary to certain tumors. Calcitonin may also be helpful in certain instances. Drugs that stop bone breakdown and reabsorption by the body, such as bisphosphonates, may be administered I.V.

## Special Considerations

Watch for hypocalcemia in patients receiving massive transfusions of citrated blood; in those with chronic diarrhea, severe infections, and insufficient dietary intake of calcium and protein (especially in elderly patients); and in those who are hyperventilating.

- ◆ Check serum calcium level every 12 to 24 hours, and report a calcium level less than 8.5 mg/dL immediately. When giving calcium supplements, frequently check the pH level because a pH lower than 7.45 inhibits calcium ionization. Check for Troussseau and Chvostek signs.
- ◆ Administer calcium gluconate slow I.V. in 5% dextrose in water or in normal saline solution. Don't add calcium gluconate I.V. to solutions containing bicarbonate; it will precipitate. When administering calcium solutions, watch for anorexia, nausea, and vomiting—possible signs of overcorrection to hypercalcemia. Never infuse more than 1 g/hour, except in an emergency. Use a volume-control device to ensure proper flow rate.
- ◆ If the patient is receiving calcium chloride, watch for abdominal discomfort.

- ◆ Monitor the patient closely for a possible drug interaction if he's receiving cardiac glycosides with large doses of oral calcium supplements; watch for signs of digoxin toxicity (anorexia, nausea, vomiting, yellow vision, and cardiac arrhythmias). Administer oral calcium supplements 1 to 1½ hours after meals or with milk.
- ◆ Provide a quiet, stress-free environment for the patient with tetany. Observe seizure precautions for patients with severe hypocalcemia.



**ALERT** *Don't confuse calcium chloride with calcium gluconate in administration; 1 g of calcium chloride has three times the calcium as 1 g of calcium gluconate.*



**PREVENTION** *To prevent hypocalcemia, advise all patients—especially elderly patients—to eat foods rich in calcium, vitamin D, and protein, such as fortified milk and cheese. Explain how important calcium is for normal bone formation and blood coagulation. Discourage chronic use of laxatives. Also, warn hypocalcemic patients not to overuse antacids, because these may aggravate the condition.*

If the patient has hypercalcemia:

- ◆ Check serum calcium level frequently. Watch for cardiac arrhythmias if serum calcium levels exceed their normal values of 8.5 to 10.5 mg/dL. Increase fluid intake to dilute calcium in serum and urine, and to prevent renal damage and dehydration. Watch for signs of heart failure in patients receiving normal saline diuresis.
- ◆ Administer loop diuretics (not thiazide diuretics), as ordered. Monitor intake and output, and check urine for renal calculi and acidity. Provide acid-ash drinks, such as cranberry or prune juice, because calcium salts are more soluble in acid than in alkali.
- ◆ Check ECG and vital signs frequently. In the patient receiving cardiac glycosides, watch for signs of toxicity, such as anorexia, nausea, vomiting, and bradycardia (often with arrhythmia).
- ◆ Ambulate the patient as soon as possible. Handle the patient with chronic hypercalcemia *gently* to prevent pathologic fractures. If the patient is bedridden, reposition the patient frequently and encourage range-of-

motion exercises to promote circulation and prevent urinary stasis and calcium loss from bone.

- ◆ To prevent recurrence, suggest a low-calcium diet, with increased fluid intake.

## CHLORIDE IMBALANCE

Hypochloremia and hyperchloremia are, respectively, conditions of deficient or excessive serum levels of the chloride anion.

### Causes and Incidence

Hypochloremia may result from:

- ◆ decreased chloride intake or absorption, as in low dietary sodium intake, sodium deficiency, potassium deficiency, metabolic alkalosis; prolonged use of mercurial diuretics; or administration of dextrose I.V. without electrolytes
- ◆ excessive chloride loss resulting from prolonged diarrhea or diaphoresis; loss of hydrochloric acid in gastric secretions due to vomiting, gastric suctioning, or gastric surgery.

Hyperchloremia may result from:

- ◆ excessive chloride intake or absorption—as in hyperingestion of ammonium chloride, or ureterointestinal anastomosis—allowing reabsorption of chloride by the bowel
- ◆ hemoconcentration from dehydration
- ◆ compensatory mechanisms for other metabolic abnormalities, as in metabolic acidosis, brainstem injury causing neurogenic hyperventilation, and hyperparathyroidism

### Pathophysiology

A predominantly extracellular anion, chloride accounts for two-thirds of all serum anions. Secreted by stomach mucosa as hydrochloric acid, it provides an acid medium that aids digestion and activation of enzymes. Chloride also participates in maintaining acid–base and body water balances, influences the osmolality or tonicity of ECF, plays a role in the exchange of oxygen and carbon dioxide in RBCs, and helps activate salivary amylase (which, in turn, activates the digestive process).

## Complications

- Depressed respirations leading to respiratory arrest (hypochloremia)
- Coma (hyperchloremia)

## Signs and Symptoms

Hypochloremia is usually associated with hyponatremia and its characteristic muscle weakness and twitching because renal chloride loss always accompanies sodium loss, and sodium reabsorption isn't possible without chloride. However, if chloride depletion results from metabolic alkalosis secondary to loss of gastric secretions, chloride is lost independently from sodium; typical symptoms are muscle hypertonicity, tetany, and shallow, depressed breathing.

Because of the natural affinity of sodium and chloride ions, hyperchloremia usually produces clinical effects associated with hypernatremia and resulting ECF volume excess (agitation, tachycardia, hypertension, pitting edema, dyspnea). Hyperchloremia associated with metabolic acidosis is due to excretion of base bicarbonate by the kidneys, and induces deep, rapid breathing; weakness; diminished cognitive ability and, ultimately, coma.

## Diagnosis

 **CONFIRMING DIAGNOSIS** A serum chloride level below 97 mEq/L confirms hypochloremia. (Supportive values in metabolic alkalosis include serum pH above 7.45 and serum carbon dioxide [CO<sub>2</sub>] level above 32 mEq/L.) A serum chloride level above 108 mEq/L confirms hyperchloremia; with metabolic acidosis, serum pH is below 7.35 and serum CO<sub>2</sub> level is below 22 mEq/L.

## Treatment

Hypochloremia therapy aims to correct the condition that causes excessive chloride loss and to give oral replacement such as salty broth. When oral therapy isn't possible, or when emergency measures are necessary, treatment may include normal saline solution I.V. (if hypovolemia is present) or chloride-containing drugs, such as ammonium chloride, to increase serum chloride levels, and potassium chloride for metabolic

alkalosis. For severe hyperchloremic acidosis, treatment consists of sodium bicarbonate I.V. to raise the serum bicarbonate level and permit renal excretion of the chloride anion, because bicarbonate and chloride compete for combination with sodium. For mild hyperchloremia, Ringer lactate solution is administered; it converts to bicarbonate in the liver, thus increasing base bicarbonate to correct acidosis.

In either kind of chloride imbalance, treatment must correct the underlying disorder.

## Special Considerations

When managing the patient with hypochloremia:

- ◆ Check serum chloride level frequently, particularly during I.V. therapy.
- ◆ Watch for signs of hyperchloremia or hypochloremia. Be alert for respiratory difficulty.
- ◆ To prevent hypochloremia, monitor laboratory results (serum electrolyte levels and blood gas values) and fluid intake and output of patients who are vulnerable to chloride imbalance, particularly those recovering from gastric surgery. Record and report excessive or continuous loss of gastric secretions. Also report prolonged infusion of dextrose in water without saline.

When managing the patient with hyperchloremia:

- ◆ Check serum electrolyte levels every 3 to 6 hours. If the patient is receiving high doses of sodium bicarbonate, watch for signs of overcorrection (metabolic alkalosis, respiratory depression) or lingering signs of hyperchloremia, which indicate inadequate treatment.



**PREVENTION** *To prevent hyperchloremia, check laboratory results for elevated serum chloride levels or potassium imbalance if the patient is receiving I.V. solutions containing sodium chloride, and monitor fluid intake and output. Also, watch for signs of metabolic acidosis. When administering I.V. fluids containing Ringer lactate solution, monitor flow rate according to the patient's age, physical condition, and bicarbonate level. Report any irregularities promptly.*

## MAGNESIUM IMBALANCE

Magnesium is the second most common cation in intracellular fluid.

Because many common foods contain magnesium, a dietary deficiency is rare. Hypomagnesemia generally follows impaired absorption, too-rapid excretion, or inadequate intake during TPN. It frequently coexists with other electrolyte imbalances, especially low calcium and potassium levels. Magnesium excess (hypermagnesemia) is common in patients with renal failure and excessive intake of magnesium-containing antacids.

## Causes and Incidence

Hypomagnesemia usually results from impaired absorption of magnesium in the intestines or excessive excretion in urine or stool. Possible causes include:

- ◆ decreased magnesium intake or absorption, as in malabsorption syndrome, chronic diarrhea, or postoperative complications after bowel resection; chronic alcoholism; prolonged diuretic therapy, NG suctioning, or administration of parenteral fluids without magnesium salts; and starvation or malnutrition
- ◆ excessive loss of magnesium, as in severe dehydration and diabetic acidosis; hyperaldosteronism and hypoparathyroidism, which result in hypokalemia and hypocalcemia; hyperparathyroidism and hypercalcemia; excessive release of adrenocortical hormones; drugs such as cisplatin and amphotericin; and diuretic therapy
- ◆ refeeding syndrome

Hypermagnesemia results from the kidneys' inability to excrete magnesium that was either absorbed from the intestines or infused. Common causes of hypermagnesemia include:

- ◆ chronic renal insufficiency
- ◆ use of laxatives (magnesium sulfate, milk of magnesia, and magnesium citrate solutions), especially with renal insufficiency
- ◆ overuse of magnesium-containing antacids
- ◆ severe dehydration (resulting oliguria can cause magnesium retention)
- ◆ overcorrection of hypomagnesemia

## Pathophysiology

Magnesium's major function is to enhance neuromuscular integration; it also stimulates PTH secretion, thus regulating intracellular fluid calcium

levels. Therefore, magnesium deficiency (hypomagnesemia) may result in transient hypoparathyroidism or interference with the peripheral action of PTH. Magnesium may also regulate skeletal muscles through its influence on calcium utilization by depressing acetylcholine release at synaptic junctions. In addition, magnesium activates many enzymes for proper carbohydrate and protein metabolism, aids in cell metabolism and the transport of sodium and potassium across cell membranes, and influences sodium, potassium, calcium, and protein levels.

About one-third of magnesium taken into the body is absorbed through the small intestine and is eventually excreted in the urine; the remaining unabsorbed magnesium is excreted in the stool.

## Complications

- ◆ Cardiac arrhythmias, hypoparathyroidism, seizures, confusion, and coma (hypomagnesemia)
- ◆ Complete heart block and respiratory paralysis (hypermagnesemia)

## Signs and Symptoms

Hypomagnesemia causes neuromuscular irritability and cardiac arrhythmias. Hypermagnesemia causes CNS and respiratory depression, in addition to neuromuscular and cardiac effects. (See *Signs and symptoms of magnesium imbalance*, page 534.)

## Signs and symptoms of magnesium imbalance

|                | <i>Dysfunction Hypomagnesemia</i>  | <i>Hypermagnesemia</i>   |
|----------------|--|--|
| Cardiovascular | ◆ Arrhythmias (such as torsades de pointes), vasomotor changes (vasodilation and hypotension) and, occasionally, hypertension              | ◆ Bradycardia, weak pulse, hypotension, heart block, cardiac arrest (common with serum levels of 25 mEq/L)                       |
| Neurologic     | ◆ Confusion, delusions, hallucinations, seizures   | ◆ Drowsiness, flushing, lethargy, confusion, diminished sensorium  |
| Neuromuscular  | ◆ Hyperirritability, tetany, leg and foot cramps, Chvostek sign (facial muscle spasms induced by tapping the branches of the facial nerve) | ◆ Diminished reflexes, muscle weakness, flaccid paralysis, respiratory muscle paralysis that may cause respiratory insufficiency |

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## Diagnosis

 **CONFIRMING DIAGNOSIS** Serum magnesium level less than 1.5 mEq/L confirms hypomagnesemia; a level greater than 2.5 mEq/L confirms hypermagnesemia.

Low levels of other serum electrolytes (especially potassium and calcium) often coexist with hypomagnesemia. In fact, unresponsiveness to correct treatment for hypokalemia strongly suggests hypomagnesemia. Similarly, elevated levels of other serum electrolytes are associated with hypermagnesemia.

## Treatment

Therapy for magnesium imbalance aims to identify and correct the underlying cause.

Treatment of mild hypomagnesemia consists of daily magnesium supplements I.M. or orally; of severe hypomagnesemia, magnesium sulfate I.V. (10 to 40 mEq/L diluted in I.V. fluid). Magnesium intoxication (a possible adverse effect) requires calcium gluconate I.V.

Therapy for hypermagnesemia includes increased fluid intake and loop diuretics (such as furosemide) with impaired renal function; calcium gluconate (10%), a magnesium antagonist, for temporary relief of symptoms in an emergency; and peritoneal dialysis or hemodialysis if renal function fails or if excess magnesium can't be eliminated.

## Special Considerations

For patients with hypomagnesemia:

- ◆ Monitor serum electrolyte levels (including magnesium, calcium, and potassium) daily for mild deficits and every 6 to 12 hours during replacement therapy.
- ◆ Measure intake and output frequently. (Urine output shouldn't fall below 0.5 to 1 mL/kg/day in patients with healthy body weight; in heavier patients, <50 mL/hour is a cause for concern.) Remember, the kidneys excrete excess magnesium, and hypermagnesemia could occur with renal insufficiency.

- ◆ Advise patients to eat foods high in magnesium, such as fish and green vegetables.
- ◆ Watch for and report signs of hypomagnesemia in patients with predisposing diseases or conditions, especially those not permitted anything by mouth or who receive I.V. fluids without magnesium.



**ALERT** Monitor vital signs during I.V. therapy. Infuse magnesium replacement slowly and watch for bradycardia, heart block, and decreased respiratory rate. Have calcium gluconate I.V. available to reverse hypermagnesemia from overcorrection. In patients with torsade de pointes, elevated magnesium levels are therapeutic.

For patients with hypermagnesemia:

- ◆ Frequently assess level of consciousness, muscle activity, and vital signs.
- ◆ Keep accurate intake and output records. Provide sufficient fluids for adequate hydration and maintenance of renal function.
- ◆ Report abnormal serum electrolyte levels immediately.
- ◆ Monitor and report ECG changes (peaked T waves, increased PR intervals, widened QRS complex).
- ◆ Watch patients receiving cardiac glycosides and calcium gluconate simultaneously, because calcium excess enhances digoxin action, predisposing the patient to digoxin toxicity.
- ◆ Advise patients, particularly elderly patients and patients with compromised renal function, not to abuse laxatives and antacids containing magnesium.
- ◆ Watch for signs of hypermagnesemia in predisposed patients. Observe closely for respiratory distress if magnesium serum levels rise above 10 mEq/L.

## PHOSPHORUS IMBALANCE

Phosphorus exists primarily in inorganic combination with calcium in teeth and bones. The incidence of hypophosphatemia varies with the underlying cause; hyperphosphatemia occurs most often in children who tend to consume more phosphorus-rich foods and beverages than adults, and in children and adults with renal insufficiency. The prognosis for both conditions depends on the underlying cause.

## Causes and Incidence

Hypophosphatemia is usually the result of inadequate dietary intake; it's often related to malnutrition resulting from a prolonged catabolic state or chronic alcoholism. It may also stem from intestinal malabsorption, chronic diarrhea, hyperparathyroidism with resultant hypercalcemia, hypomagnesemia, or deficiency of vitamin D, which is necessary for intestinal phosphorus absorption. Other causes include chronic use of antacids containing aluminum hydroxide, use of parenteral nutrition solution with inadequate phosphate content, renal tubular defects, tissue damage in which phosphorus is released by injured cells, refeeding syndrome, and diabetic acidosis.

Hyperphosphatemia is generally secondary to hypocalcemia, hypervitaminosis D, hypoparathyroidism, or renal failure (often due to stress or injury). It may also result from overuse of laxatives with phosphates or phosphate enemas.

## Pathophysiology

In ECF, the phosphate ion supports several metabolic functions: utilization of B vitamins, acid–base homeostasis, bone formation, nerve and muscle activity, cell division, transmission of hereditary traits, and metabolism of carbohydrates, proteins, and fats. Renal tubular reabsorption of phosphate is inversely regulated by calcium levels—an increase in phosphorus causes a decrease in calcium. An imbalance causes hypophosphatemia or hyperphosphatemia.

## Complications

- ◆ *Hypophosphatemia*—heart failure, shock, arrhythmias, rhabdomyolysis, seizures, and coma
- ◆ *Hyperphosphatemia*—soft-tissue complications

## Signs and Symptoms

Hypophosphatemia produces anorexia, muscle weakness, tremor, paresthesia and, when persistent, osteomalacia, causing bone pain. Impaired RBC functions may occur in hypophosphatemia due to alterations in oxyhemoglobin dissociation, which may result in peripheral hypoxia.

Hyperphosphatemia usually remains asymptomatic unless it results in hypocalcemia, with tetany and seizures.

## Diagnosis

 **CONFIRMING DIAGNOSIS** Serum phosphorus levels less than 1.7 mEq/L or 2.5 mg/dL confirm hypophosphatemia. Urine phosphorus levels above 1.3 g/24 hours support this diagnosis. Serum phosphorus levels above 2.6 mEq/L or 4.5 mg/dL confirm hyperphosphatemia. Supportive values include decreased levels of serum calcium (<9 mg/dL) and urine phosphorus (<0.9 g/24 hours).

## Treatment

Treatment aims to correct the underlying cause of phosphorus imbalance. Until this is done, the management of hypophosphatemia consists of phosphorus replacement with a high-phosphorus diet and oral administration of phosphate salt tablets or capsules. (See *Foods high in phosphorus*.) Severe hypophosphatemia requires I.V. infusion of potassium phosphate. Severe hyperphosphatemia may require peritoneal dialysis or hemodialysis to lower the serum phosphorus level.

### Foods high in phosphorus

| <b>Food</b>               | <b>Portion</b> | <b>Amount (mg)</b> |
|---------------------------|----------------|--------------------|
| Almonds                   | 1 oz           | 134                |
| Beef                      | 3 oz           | 173                |
| Egg                       | 1 large        | 104                |
| Carbonated cola beverages | 12 oz          | 40                 |
| Milk (skim)               | 8 oz           | 247                |
| Turkey (roasted)          | 3 oz           | 173                |

### Special Considerations

- ♦ Carefully monitor serum electrolyte, calcium, magnesium, and phosphorus levels. Report any changes immediately.

To manage hypophosphatemia:

- ◆ Record intake and output accurately. Administer potassium phosphate via slow I.V. to prevent overcorrection to hyperphosphatemia. Assess renal function and be alert for hypocalcemia when giving phosphate supplements. If phosphate salt tablets cause nausea, use capsules instead.
- ◆ To prevent recurrence, advise the patient to follow a high-phosphorus diet containing milk and milk products, kidney, liver, turkey, and dried fruits.

To manage hyperphosphatemia:

- ◆ Monitor intake and output. If urine output falls below 25 mL/hour or 600 mL/day, notify the physician immediately, because decreased output can seriously affect renal clearance of excess serum phosphorus.
- ◆ Watch for signs of hypocalcemia, such as muscle twitching and tetany, which often accompany hyperphosphatemia.
- ◆ To prevent recurrence, advise the patient to eat foods with low phosphorus content such as vegetables. Obtain dietary consultation if the condition results from chronic renal insufficiency.

## **SYNDROME OF INAPPROPRIATE ANTIDIURETIC HORMONE**

SIADH, also known as *dilutional hyponatremia*. The prognosis depends on the underlying disorder and response to treatment.

### **Causes and Incidence**

The most common cause of SIADH (80% of patients) is oat cell carcinoma of the lung, which secretes excessive ADH or vasopressor-like substances. Other neoplastic diseases, such as pancreatic and prostatic cancer, Hodgkin lymphoma, and thymoma, may also trigger SIADH.

Less common causes include:

- ◆ CNS disorders: brain tumor or abscess, stroke, head injury, Guillain–Barré syndrome, and lupus erythematosus
- ◆ pulmonary disorders: pneumonia, tuberculosis, lung abscess, and positive-pressure ventilation
- ◆ drugs: chlorpropamide, vincristine, cyclophosphamide, carbamazepine, clofibrate, and morphine
- ◆ miscellaneous conditions: myxedema and psychosis

### **Pathophysiology**

SIADH is marked by excessive release of ADH, which disturbs fluid and electrolyte balance. Such disturbances result from the inability to excrete dilute urine, free water retention, ECF volume expansion, and hyponatremia. SIADH occurs secondary to diseases that affect the osmoreceptors (supraoptic nucleus) of the hypothalamus.

## Complications

- ◆ Water intoxication
- ◆ Cerebral edema
- ◆ Severe hyponatremia
- ◆ Coma

## Signs and Symptoms

SIADH may produce weight gain despite anorexia, nausea, and vomiting; muscle weakness; restlessness; and, possibly, coma and seizures. Edema is rare unless water overload exceeds 4 L because much of the free water excess is within cellular boundaries.

## Diagnosis

A complete medical history revealing positive water balance may suggest SIADH.

 **CONFIRMING DIAGNOSIS** Serum osmolality less than 280 mOsm/kg of water and a serum sodium level below 123 mEq/L confirm the diagnosis (normal urine osmolality is 1½ times serum values).

Supportive laboratory values include high urine sodium secretion ( $>20$  mEq/L) without diuretics and high urine osmolality. In addition, diagnostic studies show normal renal function and no evidence of dehydration.

## Treatment

Treatment for SIADH is symptomatic and begins with restricted water intake (500 to 1,000 mL/day). For chronic treatment, fluid restriction, vasopressin receptor antagonist, loop diuretics, increased salt intake, urea, mannitol, and demeclocycline.

With acute severe water intoxication, fluid restriction, vasopressin receptor antagonist, furosemide, hypertonic saline to replace excreted

sodium.

When possible, treatment should include correction of the underlying cause of SIADH. If SIADH is due to cancer, success in alleviating water retention may be obtained by surgical resection, irradiation, or chemotherapy.

## Special Considerations

- ◆ Closely monitor and record intake and output, vital signs, and daily weight. Watch for hyponatremia.
- ◆ Observe the patient for restlessness, irritability, seizures, heart failure, and unresponsiveness due to hyponatremia and water intoxication.
- ◆ To prevent water intoxication, explain to the patient and family why they must restrict intake.

## METABOLIC ACIDOSIS

Symptoms result from the body's attempts to correct the acidotic condition through compensatory mechanisms in the lungs, kidneys, and cells. Metabolic acidosis is more prevalent among children, who are vulnerable to acid–base imbalance because their metabolic rates are faster and their ratios of water to total body weight are lower. Severe or untreated metabolic acidosis can be fatal.

### Causes and Incidence

Metabolic acidosis usually results from excessive fat burning in the absence of usable carbohydrates. This can be caused by diabetic ketoacidosis, chronic alcoholism, malnutrition, or a low-carbohydrate, high-fat diet—all of which produce more keto acids than the metabolic process can handle. Other causes include:

- ◆ anaerobic carbohydrate metabolism: a decrease in tissue oxygenation or perfusion (as occurs with pump failure after myocardial infarction, or with pulmonary or hepatic disease, shock, or anemia) forces a shift from aerobic to anaerobic metabolism, causing a corresponding rise in lactic acid level
- ◆ renal insufficiency and failure (renal acidosis): underexcretion of metabolized acids or inability to conserve base

- ◆ diarrhea and intestinal malabsorption: loss of sodium bicarbonate from the intestines, causing the bicarbonate buffer system to shift to the acidic side. For example, ureteroenterostomy and Crohn disease can also induce metabolic acidosis.

Less often, metabolic acidosis results from salicylate intoxication (overuse of aspirin), exogenous poisoning, or Addison disease with an increased excretion of sodium and chloride, and retention of potassium ions.

## Pathophysiology

Metabolic acidosis is a physiologic state of excess acid accumulation and deficient base bicarbonate produced by an underlying pathologic disorder.

## Complications

- ◆ Coma
- ◆ Arrhythmias
- ◆ Cardiac arrest

## Signs and Symptoms

In mild acidosis, the underlying disease's symptoms may obscure any direct clinical evidence. Metabolic acidosis typically begins with headache and lethargy, progressing to drowsiness, CNS depression, Kussmaul respirations (as the lungs attempt to compensate by “blowing off” carbon dioxide), stupor and, if the condition is severe and goes untreated, coma and death. Associated GI distress usually produces anorexia, nausea, vomiting, and diarrhea, and may lead to dehydration. Underlying diabetes mellitus may cause fruity breath from catabolism of fats and excretion of accumulated acetone through the lungs.

## Diagnosis

 **CONFIRMING DIAGNOSIS** Arterial pH below 7.35 confirms metabolic acidosis. In severe acidotic states, pH may fall to 7.10, and the partial pressure of arterial carbon dioxide may be normal or below 34 mm Hg as compensatory mechanisms take hold. Bicarbonate may be below 22 mEq/L.

A metabolic panel can help reveal the cause and severity of metabolic acidosis. A complete blood count can be done to help assess possible causes as well. Supportive findings include:

- ◆ urine pH: below 4.5 in the absence of renal disease
- ◆ serum potassium levels: above 5.5 mEq/L from chemical buffering
- ◆ glucose levels: above 150 mg/dL in diabetes
- ◆ serum ketone bodies: elevated levels in diabetes mellitus
- ◆ serum osmolarity: increased levels, as in hyperosmolar hyperglycemic nonketotic acidosis or dehydration
- ◆ plasma lactic acid: elevated levels in lactic acidosis
- ◆ anion gap: greater than 14 mEq/L indicating metabolic acidosis (diabetic ketoacidosis, aspirin overdose, alcohol poisoning) (See *Anion gap*.)

## Anion Gap

The anion gap is the difference between concentrations of serum cations and anions—determined by measuring one cation (sodium) and two anions (chloride and bicarbonate). The normal concentration of sodium is 140 mEq/L; of chloride, 102 mEq/L; and of bicarbonate, 26 mEq/L. Thus, the anion gap between *measured* cations (actually sodium alone) and *measured* anions is about 12 mEq/L (140 minus 128).

Concentrations of potassium, calcium, and magnesium (*unmeasured* cations), or proteins, phosphate, sulfate, and organic acids (*unmeasured* anions) aren't needed to measure the anion gap. Added together, the concentration of unmeasured cations would be about 11 mEq/L; of unmeasured anions, about 23 mEq/L. Thus, the normal anion gap between unmeasured cations and anions is about 12 mEq/L (23 minus 11)—plus or minus 2 mEq/L for normal variation. An anion gap over 14 mEq/L indicates *metabolic acidosis*. It may result from accumulation of excess organic acids or from retention of hydrogen ions, which chemically bond with bicarbonate and decrease bicarbonate levels.

## Treatment

In metabolic acidosis, treatment consists of administration of sodium bicarbonate I.V. for severe cases, evaluation and correction of electrolyte

imbalances and, ultimately, correction of the underlying cause. For example, in diabetic ketoacidosis, a low-dose continuous I.V. infusion of insulin is recommended.

## Special Considerations

- ◆ Keep sodium bicarbonate ampules handy for emergency administration. Monitor vital signs, laboratory results, and level of consciousness frequently because changes can occur rapidly.
- ◆ In diabetic acidosis, watch for secondary changes due to hypovolemia, such as decreasing blood pressure.
- ◆ Record intake and output accurately to monitor renal function. Watch for signs of excessive serum potassium—weakness, flaccid paralysis, and arrhythmias, possibly leading to cardiac arrest. After treatment, check for overcorrection to hypokalemia.
- ◆ Because metabolic acidosis commonly causes vomiting, position the patient to prevent aspiration. Prepare for possible seizures with seizure precautions.
- ◆ Provide good oral hygiene. Use sodium bicarbonate washes to neutralize mouth acids, and lubricate the patient's lips with lemon and glycerin swabs as indicated.



**PREVENTION** Carefully observe patients receiving I.V. therapy or who have intestinal tubes in place as well as those suffering from shock, hyperthyroidism, hepatic disease, circulatory failure, or dehydration. Teach the patient with diabetes how to routinely test urine for glucose and acetone, and encourage strict adherence to insulin or oral hypoglycemic therapy.

## METABOLIC ALKALOSIS

With early diagnosis and prompt treatment, prognosis is good; however, untreated metabolic alkalosis may lead to coma and death.

### Causes and Incidence

Metabolic alkalosis results from loss of acid, retention of base, or renal mechanisms associated with decreased serum levels of potassium and chloride.

Causes of critical acid loss include vomiting, NG tube drainage or lavage without adequate electrolyte replacement, fistulas, and the use of steroids and certain diuretics (furosemide, thiazides, and ethacrynic acid). Hyperadrenocorticism is another cause of severe acid loss. Cushing disease, primary hyperaldosteronism, and Bartter syndrome, for example, all lead to retention of sodium and chloride, and urinary loss of potassium and hydrogen.

Excessive base retention can result from excessive intake of bicarbonate of soda or other antacids (usually for treatment of gastritis or peptic ulcer), excessive intake of absorbable alkali (as in milk-alkali syndrome, often seen in patients with peptic ulcers), administration of excessive amounts of I.V. fluids with high concentrations of bicarbonate or lactate, or respiratory insufficiency—all of which cause chronic hypercapnia from high levels of plasma bicarbonate.

## **Pathophysiology**

A clinical state marked by decreased amounts of acid or increased amounts of base bicarbonate, metabolic alkalosis causes metabolic, respiratory, and renal responses, producing characteristic symptoms (most notably hypoventilation). This condition is always secondary to an underlying cause.

## **Complications**

- ◆ Coma
- ◆ Atrioventricular arrhythmias

## **Signs and Symptoms**

Clinical features of metabolic alkalosis result from the body's attempt to correct the acid–base imbalance, primarily through hypoventilation. Other manifestations include irritability, picking at bedclothes (carphology), twitching, confusion, nausea, vomiting, and diarrhea (which aggravates alkalosis). Cardiovascular abnormalities (such as atrial tachycardia) and respiratory disturbances (such as cyanosis and apnea) also occur. In the alkalotic patient, diminished peripheral blood flow during repeated blood pressure checks may provoke carpopedal spasm in the hand—a possible sign of impending tetany (Trousseau sign). Uncorrected metabolic alkalosis may progress to seizures and coma.

## Diagnosis

 **CONFIRMING DIAGNOSIS** Blood pH level greater than 7.45 and bicarbonate levels above 29 mEq/L confirm the diagnosis. A partial pressure of carbon dioxide above 45 mm Hg indicates attempts at respiratory compensation. Serum electrolyte studies show low potassium, calcium, and chloride levels.

Other characteristic findings include:

- ◆ Urine pH is usually about 7.0.
- ◆ Urinalysis reveals alkalinity after the renal compensatory mechanism begins to excrete bicarbonate.
- ◆ ECG may show low T wave, merging with a U wave (secondary to hypocalcemia from metabolic alkalosis), and atrial or sinus tachycardia.

## Treatment

Treatment aims to correct the underlying cause of metabolic alkalosis. Therapy for severe alkalosis may include cautious administration of ammonium chloride I.V. or hydrochloric acid to release hydrogen chloride and restore concentration of ECF and chloride levels. Potassium chloride and normal saline solution (except in the presence of heart failure) are usually sufficient to replace losses from gastric drainage. Electrolyte replacement with potassium chloride and discontinuing diuretics correct metabolic alkalosis resulting from potent diuretic therapy.

Oral or I.V. acetazolamide, which enhances renal bicarbonate excretion, may be prescribed to correct metabolic alkalosis without rapid volume expansion. Because acetazolamide also enhances potassium excretion, potassium may have to be administered before giving this drug.

## Special Considerations

Structure the care plan around cautious I.V. therapy, keen observation, and strict monitoring of the patient's status.

- ◆ Dilute potassium when giving I.V. containing potassium salts. Monitor the infusion rate to prevent damage to blood vessels; watch for signs of phlebitis. When administering ammonium chloride 0.9%, limit the infusion rate to 1 L in 4 hours; faster administration may cause

hemolysis of RBCs. Avoid overdosage because it may cause overcorrection to metabolic acidosis. Don't give ammonium chloride to patients with signs of hepatic or renal disease; instead, use hydrochloric acid.

- ◆ Watch closely for signs of muscle weakness, tetany, or decreased activity. Monitor vital signs frequently and record intake and output to evaluate respiratory, fluid, and electrolyte status. Remember, respiratory rate usually decreases in an effort to compensate for alkalosis. Hypotension and tachycardia may indicate electrolyte imbalance, especially hypokalemia.
- ◆ Observe seizure precautions.



**PREVENTION** To prevent metabolic alkalosis, warn patients against overusing alkaline agents. Irrigate NG tubes with isotonic saline solution instead of plain water to prevent loss of gastric electrolytes. Monitor I.V. fluid concentrations of bicarbonate or lactate. Teach patients with ulcers to recognize signs of milk-alkali syndrome: a distaste for milk, anorexia, weakness, and lethargy.

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# 12

## Eye Disorders

### **Introduction**

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Vision, The Most Complex Sense, Has Been The Focus Of Significant Medical And Surgical Innovations. Disorders That Affect The Eye Can Lead To Vision Loss Or Impairment; Routine Ophthalmic Examinations And Early Treatment Can Help Prevent It.

### **REVIEW OF ANATOMY**

The visual system consists mainly of the eyeball, optic nerves, extraocular muscles, cranial nerves, blood vessels, orbital fat, and lacrimal system, which are all housed within the bony orbit, and the eyelid, which covers the eye, moistens it, and protects it from injury.

The orbit (also called the *socket*) encloses the eye in a protective recess in the skull. Its seven bones—frontal, sphenoid, zygomatic, maxillary, palatine, ethmoid, and lacrimal—form a cone. The apex of this cone points toward the brain, and the cone’s base forms the orbital rim.

Extraocular muscles hold the eyes in place and control their movement, as described below:

- ◆ *superior rectus*: elevates the eye upward; adducts and rotates the eye inward

- ◆ *inferior rectus*: depresses the eye downward; adducts and rotates the eye outward
- ◆ *lateral rectus*: abducts or turns the eye outward (laterally)
- ◆ *medial rectus*: adducts or turns the eye inward (medially)
- ◆ *superior oblique*: rotates the eye inward; abducts and depresses the eye
- ◆ *inferior oblique*: rotates the eye outward; abducts and elevates the eye

The actions of these muscles are mutually antagonistic: As one contracts, its opposing muscle relaxes.



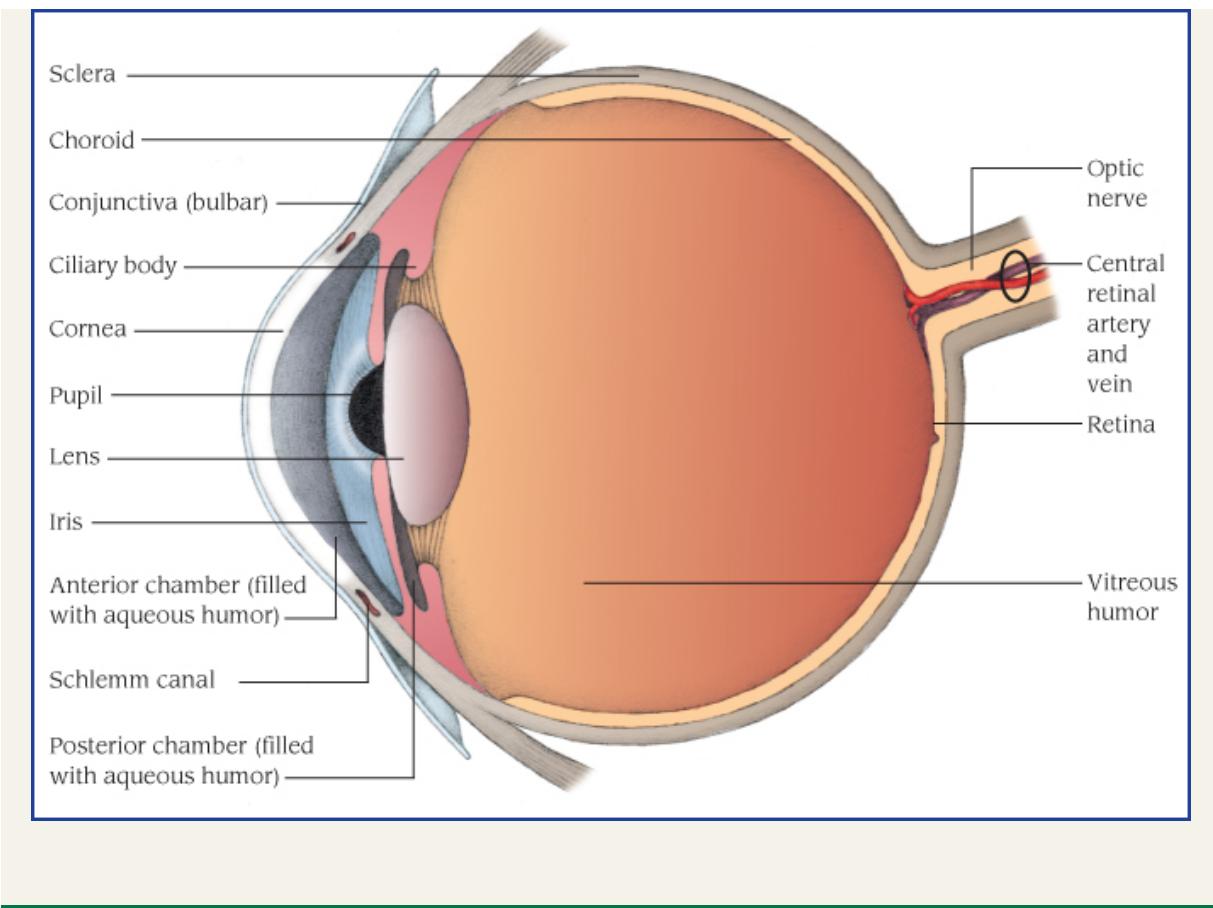
**ELDER TIP** Eye structure and activity change with age. The eyes set deeper in their sockets and the eyelids lose their elasticity, allowing the orbital fat to protrude forward. Eyelids appear more saggy and wrinkled.

## OCULAR LAYERS

The Eye Has Three Structural Layers: The Sclera And Cornea, The Uveal Tract, And The Retina. (See *Cross Section Of The Eye*, Page 595.)



## PATHOPHYSIOLOGY CROSS SECTION OF THE EYE



The sclera is the dense, white, fibrous outer protective coat of the eye. It meets the cornea at the limbus (corneoscleral junction) anteriorly and the dural sheath of the optic nerve posteriorly. The lamina cribrosa is a sievelike structure composed of a few strands of scleral tissue through which the optic nerve bundles pass. The sclera is covered by the episclera, a thin layer of fine elastic tissue.



**ELDER TIP** In older adults, lens changes occur typically with the formation of a cataract. The vitreous body liquefies and pulls away from the retina, generating floating vitreous debris and vitreous detachments.

The cornea is the transparent, avascular, dome-shaped layer of the eye that's continuous with the sclera. The cornea consists of five layers: the epithelium, which contains sensory nerves; Bowman's membrane, the basement membrane for the epithelial cells; the stroma, or supporting tissue (90% of the corneal structure); Descemet's membrane, containing many elastic fibers; and the endothelium, a single layer of cells that acts as a

pump to maintain proper dehydration or detumescence of the cornea. Aqueous humor bathes the posterior surface of the cornea, providing it with nutrients, and maintaining intraocular pressure (IOP) by volume and rate of outflow. The anterior cornea is kept moist by the tear film.



**ELDER TIP** *Dry Eye Syndrome Is More Common In The Elderly.*

*Certain Medications, Such As Histamines, Some Antidepressants, And Antihypertensives, As Well As Systemic Conditions (Such As Menopause, Diabetes, And Thyroid Disorders), Can Exacerbate Dry Eyes, Which May Result In Decreased Vision, Eye Redness, And Discomfort.*

The Middle Layer Of The Eye, The Uveal Tract, Is Pigmented And Vascular. It Consists Of The Iris And The Ciliary Body In The Anterior Portion And The Choroid In The Posterior Portion. In The Center Of The Iris Is The Pupil. The Sphincter And Dilator Muscles Control The Amount Of Light That Enters The Eye By Changing The Size Of The Pupil.



**ELDER TIP** *It is believed that atrophy of the dilator muscle fibers and increased rigidity of the blood vessels of the iris reduce pupil size, decreasing the amount of light that reaches the retina. Consequently, higher levels of illumination may be needed to improve uncorrected visual acuity in the older adult.*

The angle formed by the anterior iris surface and the posterior corneal structures contains the many minute collecting channels of the trabecular meshwork. Aqueous humor drains through these channels into an encircling venous system called *the canal of Schlemm*.

The ciliary body, which extends from the root of the iris to the peripheral retina, produces aqueous humor and controls lens accommodation through its action on the zonular fibers. The choroid, the largest part of the uveal tract, is made up of blood vessels bound externally by the suprachoroid and internally by the retina.

The retina is the innermost coat of the eye. It receives visual images in the form of light and converts the images into neural impulses. The photoreceptor cells (rods and cones) are the light-sensitive cells. These and other retina cell types are interconnected by synapses and organized in layers that transmit the neural signals to the brain. Although both rods and cones are light receptors, they respond to light differently. Rods, scattered

throughout the retina, respond to low levels of light and detect moving objects; cones, located in the fovea centralis, function best in brighter light and perceive finer details.

Three types of cones contain different visual pigments and react to specific light wavelengths: one type reacts to red light, one to green, and one to blue-violet. The eye mixes these colors into various shades.



**ELDER TIP** Many elderly patients lose their ability to discriminate blue-greens, and white objects appear yellowish; these patients may also have difficulty discriminating among pastels, violets, and yellow-greens. The retinal pigmented epithelium layer, which is just posterior to the photoreceptors, has multiple support functions, including phagocytosis of photoreceptor segments, vitamin A metabolism, and regulation of molecule transport to the retina.

## THE LENS AND ACCOMMODATION

The lens of the eye is biconvex, avascular, and transparent; the lens capsule is a semipermeable membrane that encloses the lens and allows water and nutrients to reach the lens cells. The lens changes shape (accommodation) for near and far vision. For near vision, the ciliary body contracts and relaxes the zonules, the lens becomes steeper, the pupil constricts, and the eyes converge; for far vision, the ciliary body relaxes, the zonules tighten, the lens becomes flatter, the eyes straighten, and the pupils dilate. The lens refines the refraction necessary to focus a clear image on the retina.



**ELDER TIP** In older adults, lens changes occur, typically with the formation of a cataract. Symptoms of a cataract include glare, decreased vision, and the need for increased illumination. The purpose of cataract surgery is to remove the opacified lens (cataract) and to sharpen vision.

The vitreous body, which is 99% water and a small amount of insoluble protein, constitutes two thirds of the eye's volume. This transparent, gelatinous body gives the eye its shape and contributes to the refraction of light rays. The vitreous is firmly attached to the peripheral retina near the ciliary body (anteriorly) and to the optic disk (posteriorly). The vitreous face contacts the lens; the vitreous gel rests against the retina.

## LACRIMAL APPARATUS AND EYELIDS

The lacrimal apparatus consists of the lacrimal glands, upper and lower canaliculi, lacrimal sac, and nasolacrimal duct. The main gland, located in a shallow fossa beneath the superior temporal orbital rim, secretes reflex tears. Small lacrimal glands throughout the conjunctiva are responsible for basal tear production. Multiple sebaceous glands in the eyelids produce an oily secretion that prevents tears from evaporating. With every blink, the eyelids direct the flow to the inner canthus, where the tears pool and then drain through a tiny opening called the *punctum*. The tears then pass through the canaliculi and lacrimal sac and down the nasolacrimal duct, which opens into the nasal cavity. The integrity of the lacrimal system is critical for moisturizing and also removing excess tears from the corneal surface.

The eyelids (palpebrae) consist of tarsal plates that are composed of dense connective tissue. The orbital septum—the fascia behind the orbicularis oculi muscle—acts as a barrier between the lids and the orbit. The levator palpebrae muscle elevates the upper lid. The eyelids contain three types of glands:

- ◆ glands of Zeis—modified sebaceous glands connected to the follicles of the eyelashes
- ◆ meibomian glands—sebaceous glands in the tarsal plates that secrete an oily substance as a tear film component (About 25 of these glands are found in the upper lid and about 20 in the lower lid.)
- ◆ Moll glands—ordinary sweat glands

The conjunctiva is the thin mucous membrane that lines the eyelids (palpebral conjunctiva), folds over at the fornix, and covers the surface of the eyeball (bulbar conjunctiva). The conjunctiva produces mucin, another component of the tear film. The ophthalmic, lacrimal, and multiple anastomoses of facial arteries supply blood to the lids. The space between the open lids is the palpebral fissure; the juncture of the upper and lower lids is the canthus. The junction near the nose is called the nasal, medial, or inner canthus; the junction on the temporal side, the lateral or external canthus.

## **OPTIC NERVE**

The optic nerve is composed of the nerve fibers (axons) that originate in the retina and synapse at the lateral geniculate nucleus in the brain.

Approximately 1 million nerve axons are contained in the optic nerve. The nerve from each eye exits the eye posteriorly and courses through the orbit to the optic canal. Both nerves meet at the optic chiasm, located intracranially near the pituitary gland. In the optic chiasm, part of the nerve fibers from one eye cross to the other side, and vice versa. From the synapse at the lateral geniculate nucleus, nerves carry visual information to the visual cortex, located in the majority of the occipital lobe of the brain. Some axons from the optic nerve synapse at other parts of the brain to regulate pupil responses, eye movements, and the sleep-wake cycle.

 **ELDER TIP** *Age-related vision changes are usually first noticed during the fifth decade of life and may include the inability to focus, narrowing of the visual field, reduced peripheral vision, and loss of iris elasticity producing decreased response to light and dark. In addition, as people age, production of any of the three tear film components may decrease, causing dry eyes.*

## DEPTH PERCEPTION

In normal binocular vision, a perceived image is projected onto the two foveae. Impulses then travel along the optic pathways to the occipital cortex, which perceives a single image. However, the cortex receives two images—each from a slightly different angle—giving the images perspective and providing depth perception.

## VISION TESTING

Several tests assess visual acuity and identify visual defects:

- ◆ Ishihara test determines color blindness by using a series of plates composed of a colored background, with a letter, number, or pattern of a contrasting color located in the center of each plate. The patient with deficient color perception can't perceive the differences in color or, consequently, the designs formed by the color contrasts.
- ◆ The Snellen chart or other eye charts evaluate visual acuity. Such charts use progressively smaller letters or symbols to determine central vision on a numerical scale. A person with normal acuity should be able to read the letters or recognize the symbols on the 20/20 line of the eye chart at a distance of 20'.

## SUBJECTIVE TESTING

Several tests accomplish objective testing of the eyes.

- ◆ B-mode ultrasonography delineates retinal tumors, detachments, and vitreous hemorrhages—even in the presence of opacities of the cornea and lens. A handheld B-scanner has simplified ultrasonic examination of the eye, making it possible to perform such studies in the eye care practitioner's office.
- ◆ The cover-uncover test assesses eye muscle misalignment or tendency toward misalignment. In this test, the patient stares at a small, fixed object—first from a distance of 20' (6.1 m) and then from 1' (0.3 m). The examiner covers the patient's eyes one at a time, noting any movement of the uncovered eye and the direction of any deviation. In exotropia the eyes are naturally deviated outward. In the cover-uncover test, the eyes recover by moving inward to focus. The reverse is true in esotropia.
- ◆ Duction test checks eye movement in all directions of gaze. While one eye is covered, the other eye follows a moving light. This test detects weakness of rotation due to muscle paralysis or structural dysfunction.
- ◆ Fluorescein angiography evaluates the blood vessels in the choroid and retina after I.V. injection of fluorescein dye; images of the dye-enhanced vasculature are recorded by rapid-sequence photographs of the fundus.
- ◆ Goldmann applanation, Tonopen tonometry, and pneumotonometry all measure IOP. After instilling a local anesthetic in the patient's eye, the examiner touches the Tonopen tonometer tip to the surface of the cornea. The IOP reading is displayed and measured in mm Hg. Applanation tonometry gauges the force required to flatten a small area of the central cornea, and is the most accurate method of measuring IOP. For this test, a patient must be seated at a slit lamp and the cornea stained with fluorescein dye before the prism of the applanation tonometer touches the cornea and the examiner adjusts the controls until the two lines form an “S.”
- ◆ Gonioscopy allows for direct visualization of the anterior chamber angle.
- ◆ The Maddox rod test assesses muscle dysfunction; it's especially useful in disclosing and measuring heterophoria (the tendency of the eyes to deviate). It can reveal horizontal, vertical, and, especially, torsional deviations.

- ◆ Ophthalmoscopy—direct ophthalmoscopy or binocular indirect ophthalmoscopy allows examination of the interior of the eye after the pupil has been dilated with a mydriatic. A light source and lenses are used by the examiner to focus on the posterior ocular structures (such as the retina and optic nerve).
- ◆ Refraction tests may be performed with or without cycloplegics. In cycloplegic refraction, eyedrops weaken the accommodative power of the ciliary muscle. Lenses placed in front of the eye direct light rays onto the retina, thus focusing the image so that it can be transmitted along the visual pathway. A retinoscope may be used in the same way by directing a beam of light through the pupil onto the retina; the light's shadow is neutralized by placing the appropriate lens in front of the eye.
- ◆ Slit-lamp biomicroscopic examination allows a well-illuminated examination of the eyelids and the anterior segment of the eyeball using a specialized microscope.
- ◆ Visual field tests assess the function of the retina, the optic nerve, and the optic pathways by recording the responses of the patient to light impulses directed to various areas of the visual field.

## Eyelid and Lacrimal Ducts

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### **BLEPHARITIS**

A common inflammation, blepharitis produces a red-rimmed appearance of the margins of the eyelids. It's frequently chronic and bilateral and can affect both upper and lower lids. Seborrheic blepharitis is characterized by formation of waxy scales on the eyelashes and eyelid margins, and symptoms of burning and foreign body sensation.

### **Pathophysiology**

Seborrheic blepharitis may be seen in conjunction with seborrhea of the scalp, eyebrows, and ears. It's common in elderly people and in people with red hair. Staphylococcal (ulcerative) blepharitis is characterized by the formation of dry scales along the inflamed lid margins, which also have ulcerated areas and may be associated with keratoconjunctivitis sicca, a dry eye syndrome. Staphylococcal blepharitis is associated with *Staphylococcus aureus* infection and is more common in females than in males. Both types

may coexist. Blepharitis tends to recur and become chronic. It can be controlled if treatment begins before the onset of ocular involvement.

Allergies and eyelash infestations with lice are less common causes of blepharitis. Blepharitis may also be associated with repeated styes and chalazion.

## Complications

- ◆ Keratitis
- ◆ Conjunctivitis
- ◆ Dry eyes

## Signs and Symptoms

Clinical features of blepharitis include itching, burning, foreign body sensation, and sticky, crusted eyelids on waking. This constant irritation results in unconscious rubbing of the eyes (causing reddened rims) or continual blinking. Other signs include waxy scales in seborrheic blepharitis; and flaky scales on lashes, loss of lashes, and ulcerated areas on lid margins in ulcerative blepharitis.

## Diagnosis

Diagnosis depends on patient history and characteristic symptoms. In staphylococcal blepharitis, culture of ulcerated lid margin shows *S. aureus*.

## Treatment

The goals of therapy are to control the disease and its underlying causes, maintain vision, and avoid secondary complications. Treatment depends on the type of blepharitis:

- ◆ blepharitis resulting from pediculosis—removal of nits (with forceps) or application of ophthalmic physostigmine or other ointment as an insecticide (This may cause pupil constriction and, possibly, headache, conjunctival irritation, and blurred vision from the film of ointment on the cornea.)
- ◆ seborrheic blepharitis—daily lid hygiene (using a mild shampoo on a damp applicator stick or a washcloth) and hot compresses to remove scales from the lid margins; also, frequent shampooing of the scalp and eyebrows

- ◆ staphylococcal blepharitis—warm compresses and an antibiotic, such as tetracycline or erythromycin eye ointment, may be used. For some patients, systemic antibiotics are indicated

## Special Considerations

- ◆ Instruct the patient to gently remove scales from the lid margins daily, with an applicator stick or a clean washcloth.
- ◆ Teach the patient the following method for applying warm compresses: First, run warm water into a clean bowl. Then, immerse a clean cloth in the water and wring it out. Place the warm cloth against the closed eyelid (be careful not to burn the skin). Hold the compress in place until it cools. Continue this procedure for 15 minutes.
- ◆ Antibiotic ophthalmic ointment should be applied after 15-minute application of warm compresses.
- ◆ Treatment for seborrheic blepharitis also requires attention to the face and scalp.

## EXOPHTHALMOS

Exophthalmos (also called *proptosis*) is the unilateral or bilateral bulging or protrusion of the eyeballs or their apparent forward displacement (with lid retraction). The prognosis depends on the underlying cause.

### Pathophysiology and Incidence

Exophthalmos commonly results from hyperthyroidism, particularly ophthalmic Graves disease, in which the eyeballs are displaced forward and the lids retract. Unilateral exophthalmos may also result from trauma (such as fracture of the ethmoid bone, which allows air from the sinus to enter the orbital tissue, displacing soft tissue and the eyeball forward). Exophthalmos may also stem from hemorrhage, varicosities, thrombosis, and edema, all of which similarly displace one or both eyeballs forward.

Other systemic and ocular causes include:

- ◆ infection—orbital cellulitis, panophthalmitis, and infection of the lacrimal gland or orbital tissues
- ◆ parasitic cysts—in surrounding tissue
- ◆ tumors and neoplastic diseases—in children, rhabdomyosarcomas, leukemia, gliomas of the optic nerve, dermoid cysts, teratomas,

metastatic neuroblastomas, and lymphoma; in adults, lacrimal gland tumors, mucoceles, cavernous hemangioma, meningiomas, metastatic carcinomas, and lymphoma

## Signs and Symptoms

The obvious sign is a bulging eyeball, commonly with diplopia, due to eyeball misalignment or extraocular muscle dysfunction. (See *Recognizing exophthalmos*.) A rim of the sclera may be visible below the upper lid as lid retraction occurs. Other symptoms depend on the cause: pain may accompany traumatic exophthalmos; a tumor may produce conjunctival hyperemia or chemosis; retraction of the upper lid predisposes to exposure keratitis. If exophthalmos is associated with cavernous sinus thrombosis, the patient may exhibit paresis of the muscles supplied by cranial nerves III, IV, and VI; limited ocular movement; and a septic-type (high) fever.

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## Recognizing Exophthalmos

This photograph shows the characteristic forward protrusion of the eyes from the orbit associated with exophthalmos.



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## Diagnosis

Exophthalmos is usually obvious on physical examination; exophthalmometer readings confirm diagnosis by showing the degree of anterior projection and asymmetry between the eyes (normal bar readings

range from 12 to 20 mm). The following diagnostic measures identify the cause:

- ◆ Computed tomography scan or magnetic resonance imaging detects swollen extraocular muscles or lesions within the orbit.
- ◆ Culture of discharge determines the infecting organism; sensitivity testing indicates appropriate antibiotic therapy.
- ◆ Biopsy of orbital tissue may be necessary if initial treatment fails.

## Treatment

Eye trauma may require cold compresses for the first 24 hours, followed by warm compresses, and prophylactic antibiotic therapy. After edema subsides, surgery may be necessary in a small percentage of cases. It is important to counsel patients with acute orbital fractures not to blow their nose, to avoid air entering the orbit, which may cause acute exophthalmos. Eye infection requires treatment with broad-spectrum antibiotics during the 24 hours preceding positive identification of the organism, followed by specific antibiotics. A patient with exophthalmos resulting from an orbital tumor may initially benefit from antibiotic or corticosteroid therapy. Eventually, surgical exploration of the orbit and excision of the tumor, enucleation, or exenteration may be necessary. Radiation and chemotherapy may be used when primary orbital tumors can't be fully excised as encapsulated lesions, such as in rhabdomyosarcoma lesions.

Treatment for Graves disease may include antithyroid drug therapy or partial or total thyroidectomy to control hyperthyroidism; initial high doses of systemic corticosteroids, such as prednisone, for optic neuropathy and, if lid retraction is severe, protective lubricants.

Surgery may include orbital decompression (removal of any of the orbital walls) if vision is threatened, followed by muscle surgery and then lid surgery (eyelid retraction repair).

## Special Considerations

- ◆ It is critical to protect the exposed cornea with lubricants to prevent corneal drying until the disease stabilizes or is corrected by surgery.

## PTOSIS

Ptosis (drooping of the upper eyelid) may be congenital or acquired, unilateral or bilateral, and constant or intermittent. Severe ptosis usually responds well to treatment; slight ptosis may require no treatment at all.

## **Pathophysiology**

Congenital ptosis is transmitted as an autosomal dominant trait or results from a congenital anomaly in which the levator muscles of the eyelids fail to develop. This condition is usually unilateral.

Acquired ptosis may result from any of the following:

- ◆ advanced age (involutional ptosis, the most common form, usually seen in older patients)
- ◆ mechanical factors that make the eyelid heavy, such as swelling caused by a foreign body on the palpebral surface of the eyelid or by edema, inflammation produced by a tumor or pseudotumor, or an extra fatty fold
- ◆ myogenic factors, such as muscular dystrophy or myasthenia gravis (in which the defect appears to be in humoral transmission at the myoneural junction)
- ◆ neurogenic (paralytic) factors from interference in innervation of the eyelid by the oculomotor nerve (cranial nerve III), most commonly due to trauma, diabetes, or carotid aneurysm (Ptosis due to oculomotor nerve damage produces a fixed, dilated pupil, divergent strabismus, and slight depression of the eyeball.)
- ◆ nutritional factors, such as thiamine deficiency in chronic alcoholism, hyperemesis gravidarum, and other malnutrition-producing states

Risk factors for ptosis include aging, diabetes, stroke, Horner syndrome, myasthenia gravis, and cancer that affects nerve or muscle response.

- ◆ In myasthenia gravis, ptosis results from fatigue and characteristically appears in the evening but is relieved by rest.

The child with unilateral ptosis that covers the pupil can develop an amblyopic eye from disuse or lack of eye stimulation. In bilateral ptosis, the child may elevate the brow in an attempt to compensate, wrinkling the forehead in an effort to raise the upper lid. Additionally, the child may tip the head backward to see.

## **Complication**

- ◆ Lazy eye in children (amblyopia)

## Signs and Symptoms



**PEDIATRIC TIP** *An infant with congenital ptosis has a smooth, flat upper eyelid, without the eyelid fold normally caused by the pull of the levator muscle; associated weakness of the superior rectus muscle isn't uncommon.*

## Diagnosis

Examination includes measurement of the position of the upper eyelid margin relative to the pupil, degree of eyelid excursion, presence or absence of lagophthalmos, Bell phenomenon, and eyelid crease. Diagnosis may also include these tests to determine any underlying cause:

- ◆ digital subtraction angiography or magnetic resonance imaging—aneurysm
- ◆ glucose tolerance test—diabetes
- ◆ ophthalmologic examination—foreign bodies
- ◆ patient history—chronic alcoholism
- ◆ Tensilon test—myasthenia gravis (in acquired ptosis with no history of trauma)

## Treatment

Slight ptosis that doesn't produce deformity or loss of vision requires no treatment. Severe ptosis that interferes with vision or is cosmetically undesirable usually necessitates reattachment of a stretched levator aponeurosis. Surgery to correct congenital ptosis is usually performed at age 3 or 4, but it may be done earlier if amblyopia is a concern. The surgical approach depends on the degree of ptosis. If surgery is contraindicated, special glasses with an attached suspended crutch on the frames may elevate the eyelid.

Effective treatment for ptosis also requires treatment for any underlying cause. For example, in patients with myasthenia gravis, neostigmine or steroids may be prescribed to increase the effect of acetylcholine and aid transmission of nerve impulses to muscles.

## Special Considerations

- ◆ After surgery to correct ptosis, watch for blood on the pressure patch. (Some surgical procedures may not require a patch.) Apply ointment to the sutures as prescribed.
- ◆ Emphasize to the patient and family the need to prevent accidental trauma to the surgical site until healing is complete (6 weeks). Suture line damage can precipitate recurrence of ptosis.

## ORBITAL CELLULITIS

Orbital cellulitis is an acute infection of the orbital tissues and eyelids that doesn't involve the eyeball. With treatment, the prognosis is good; if untreated, the infection may spread intracranially to the cavernous sinus or the meninges, where it can be life-threatening.

### Pathophysiology

Orbital cellulitis may result from bacterial, fungal, or parasitic infection. It can develop from direct inoculation, via the bloodstream, or spread from adjacent structures (e.g., the sinuses or eyelids). Periorbital tissues may be inoculated as a result of surgery, foreign body trauma, and even animal or insect bites.



**PEDIATRIC TIP** *The most common pathogens in children are Haemophilus influenzae, Streptococcus pneumoniae, and S. aureus. In young children, infection spreads from adjacent sinuses (especially the ethmoid air cells) and accounts for the majority of postseptal cellulitis cases. The incidence has decreased because of the use of the H. influenzae b (Hib) vaccine.*

Immunosuppressed patients are also susceptible.

### Complications

- ◆ Cavernous sinus thrombosis
- ◆ Hearing loss
- ◆ Septicemia
- ◆ Meningitis
- ◆ Optic nerve damage

## **Signs and Symptoms**

Orbital cellulitis generally produces unilateral eyelid edema, reddened eyelids, and matted lashes. Although the eyeball is initially unaffected, proptosis develops later (because of edematous tissues within the bony confines of the orbit). Other indications include extreme orbital pain, impaired eye movement, chemosis, purulent discharge from indurated areas, decreased vision, and an afferent pupillary defect. The severity of associated systemic symptoms (chills, fever, and malaise) varies according to the cause.

Complications include posterior extension, causing cavernous sinus thrombosis, panophthalmitis, meningitis, or brain abscess and, rarely, atrophy and subsequent loss of vision secondary to optic neuritis.

## **Diagnosis**

Typical clinical features establish diagnosis. Computed tomography scan or magnetic resonance imaging of the sinuses and orbit tissues will determine if the cause of the cellulitis is preseptal or if deeper structures are involved, or if a tumor is the cause of swelling. Usually the patient will also be febrile with this type of infection. Wound culture and sensitivity testing determine the causative organism and specific antibiotic therapy. Other tests include white blood cell count and ophthalmologic examination.

## **Treatment**

Prompt treatment is necessary to prevent complications. Primary treatment consists of antibiotic therapy. Systemic antibiotics (I.V. or oral) and eyedrops or ointment will be ordered. Supportive therapy consists of fluids; warm, moist compresses; and bed rest. The patient should be monitored closely. If during the initial 48 to 72 hours of treatment no improvement is seen, adjustment of antibiotics guided by drug sensitivity should be considered. If an orbital abscess is present, surgical incision and drainage may be necessary.

## **Special Considerations**

- ◆ Monitor vital signs at least every 4 hours, and maintain fluid and electrolyte balance.

- ◆ Have the patient instill antibiotic eyedrops frequently during the day and apply ointment at night.
- ◆ Apply compresses every 3 to 4 hours to localize inflammation and relieve discomfort. Teach the patient to apply these compresses. Give pain medication, as ordered, after assessing pain level.
- ◆ Before discharge, stress the importance of completing prescribed antibiotic therapy. To prevent orbital cellulitis, tell the patient to maintain good general hygiene and to carefully clean abrasions and cuts that occur near the orbit.
- ◆ Ensure patient has appropriate follow-up.



## PREVENTION

- ◆ Use *Hib* vaccination to prevent *Haemophilus* infection in children.
- ◆ Treat sinus and dental infections early to decrease spread to the eye.

## DACRYOCYSTITIS

Dacryocystitis is an infection of the lacrimal sac. It can be acute, chronic, or congenital. In infants, dacryocystitis results from congenital atresia of the nasolacrimal duct; in adults, it results from an obstruction (dacyostenosis) of the nasolacrimal duct (most common in women older than age 40).

### Pathophysiology

Atresia of the nasolacrimal ducts results from failure of canalization or, in the first few months of life, from blockage when the membrane that separates the lower part of the nasolacrimal duct and the inferior nasal meatus fails to open spontaneously before tear secretion. Bony obstruction of the duct may also occur.

In acute dacryocystitis, *S. aureus* and, occasionally, beta-hemolytic streptococci are the cause. In chronic dacryocystitis, *S. pneumoniae* or, sometimes, a fungus—such as *Actinomyces* or *Candida albicans*—is the causative organism. Primary lumps and secondary tumors from sinuses, nose, and orbits have also been reported as causes.

### Complication

- ◆ Orbital cellulitis

## **Signs and Symptoms**

The hallmark of both the acute and chronic forms of dacryocystitis is constant tearing. Other symptoms of dacryocystitis include inflammation and tenderness over the nasolacrimal sac; pressure over this area may fail to produce purulent discharge from the punctum. Acute dacryocystitis is painful for the patient.

## **Diagnosis**

Clinical features and a physical examination suggest dacryocystitis. Culture of the discharged material demonstrates *S. aureus* and, occasionally, beta-hemolytic streptococci in acute dacryocystitis, and *S. pneumoniae* or *C. albicans* in the chronic form. The white blood cell count may be elevated in the acute form; in the chronic form, it's generally normal. An X-ray after injection of a radiopaque medium (dacryocystography) locates the atresia in infants.

## **Treatment**

Treatment for acute dacryocystitis consists of warm compresses, topical and systemic antibiotic therapy, and, occasionally, incision and drainage. Chronic dacryocystitis may eventually require dacryocystorhinostomy. Laser-assisted endoscopic dacryocystorhinostomy and balloon dilatation or probing of the nasolacrimal system may also be used.

Therapy for nasolacrimal duct obstruction in an infant consists of careful massage of the area over the lacrimal sac four times a day for 6 to 9 months. If this fails to open the duct, dilation of the punctum and probing of the duct are necessary.

## **Special Considerations**

- ◆ Check the patient history for possible allergy to antibiotics before administration. Emphasize the importance of precise compliance with the prescribed antibiotic regimen.
- ◆ Tell the adult patient what to expect after surgery. The patient should expect to have ice compresses over the surgical site and will have bruising and swelling.
- ◆ Monitor blood loss by counting dressings used to collect the blood.

- ◆ Apply ice compresses postoperatively. A small adhesive bandage may be placed over the suture line to protect it from damage.

## CHALAZION

A chalazion is a chronic granulomatous inflammation of a meibomian gland or gland of Zeis in the upper or lower eyelid. (There are ~100 of these glands located near the eyelashes.) This common eye disorder is characterized by localized swelling within the tarsal plate, or it may break through the conjunctival or skin side. Mild irritation and blurred vision usually develop slowly over several weeks. (See *Recognizing chalazion*.) A chalazion may become large enough to press on the eyeball, causing astigmatism. A large chalazion seldom subsides spontaneously. It's generally benign and chronic, and can occur at any age. In some patients, it's apt to recur.

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### Recognizing Chalazion

A chalazion is a nontender granulomatous inflammation of a meibomian gland on the upper or lower eyelid.



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### Pathophysiology

Obstruction of the meibomian (sebaceous) gland duct causes a chalazion.

### Complication

- ◆ Astigmatism

## Signs and Symptoms

A chalazion occurs as a painless, hard lump that usually points toward the conjunctival side of the eyelid. Eversion of the lid reveals a red or red-yellow elevated area on the conjunctival surface. Otherwise, it's seen as an indurated bump under the skin of the upper eyelid.

## Diagnosis

Diagnosis requires visual examination and palpation of the eyelid, revealing a small bump or nodule. Persistently recurrent chalazions, especially in an adult, necessitate biopsy to rule out sebaceous cell carcinoma.

## Treatment

Initial treatment consists of application of warm compresses for 10 to 15 minutes at least four times a day to open the lumen of the gland, soften the hardened oils blocking the duct, and promote drainage and healing. If such therapy fails, or if the chalazion presses on the eyeball or causes a severe cosmetic problem, steroid injection or incision and curettage under local anesthetic may be necessary. After such surgery, a pressure eye patch applied for 4 to 6 hours controls bleeding and swelling. After removal of the patch, treatment again consists of warm compresses. Antibiotic eyedrops are occasionally prescribed before and after cyst removal, but otherwise are of little value.

## Special Considerations

- ◆ Instruct the patient how to properly apply warm compresses: Educate the patient to take special care to avoid burning the skin, to always use a clean cloth, and to discard used compresses. In addition, instruct the patient to start applying warm compresses at the first sign of lid irritation to increase the blood supply and keep the lumen open.
- ◆ Teach the patient how to instill antibiotic eyedrops.

## STYE

A localized, purulent staphylococcal infection, a stye (or hordeolum) can occur externally (in the lumen of the smaller glands of Zeis or in Moll

glands) or internally (in the larger meibomian gland). A stye can occur at any age. Generally, styes are self-limiting and respond well to hot, moist compresses. More than one may occur at the same time. If untreated, a stye can eventually lead to cellulitis of the eyelid. Styes can also develop into a chalazion if gland ducts are fully blocked.

## Pathophysiology

Skin bacteria that enter eyelash hair follicles and cause inflammation can result in stye formation. Risk factors include blepharitis, diabetes and other chronic debilitating illnesses, and seborrhea.

## Complication

- ◆ Cellulitis of the eyelid

## Signs and Symptoms

Typically, a stye produces redness, swelling, and pain. An abscess frequently forms at the lid margin, with an eyelash pointing outward from its center. (See *Recognizing a stye*.)

### Recognizing A Stye

A stye is a localized red, swollen, and tender abscess of the lid glands.



## Diagnosis

Visual examination generally confirms this infection. Culture of purulent material from the abscess usually reveals a staphylococcal organism.

## Treatment

Treatment consists of warm compresses applied for 10 to 15 minutes, four times a day for 3 to 4 days, to facilitate drainage of the abscess, to relieve pain and inflammation, and to promote suppuration. Drug therapy includes antibiotic eyedrops or ointment and, occasionally, a systemic antibiotic for secondary eyelid cellulitis. If conservative treatment fails, incision and drainage may be necessary.

## Special Considerations

- ◆ Instruct the patient to use a clean cloth for each application of warm compresses and to dispose of it or launder it separately.
- ◆ Warn against squeezing the stye; this spreads the infection and may cause cellulitis.
- ◆ Teach the patient or family members the proper technique for instilling eyedrops or ointments into the cul-de-sac of the lower eyelid.



**PREVENTION** Teach proper eye hygiene, such as washing hands and using clean towels, to prevent recurrent infections.

## Conjunctival Disorders

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### INCLUSION CONJUNCTIVITIS

Inclusion conjunctivitis is an acute ocular inflammation resulting from infection by *Chlamydia trachomatis*. Although inclusion conjunctivitis occasionally becomes chronic, the prognosis is usually good.

### Pathophysiology

*Chlamydia trachomatis* is an obligate intracellular organism of the lymphogranuloma venereum serotype group. Serotypes D through K are sexually transmitted, and secondary eye involvement in adults occurs in about 1 in 300 genital cases. Because contaminated cervical secretions infect the eyes of the neonate during birth, inclusion conjunctivitis is an

important cause of ophthalmia neonatorum. Ocular chlamydial disease occurs most frequently in adults between ages 18 and 30.

## Complications

- ◆ Otitis media
- ◆ Blindness

## Signs and Symptoms

Inclusion conjunctivitis develops 5 to 12 days after contamination (it takes longer to develop than gonococcal ophthalmia). In a neonate, reddened eyelids and tearing with moderate mucoid discharge are presenting symptoms. In neonates, pseudo membranes may form, which can lead to conjunctival scarring. In adults, follicles appear inside the lower eyelids; such follicles don't form in infants because the lymphoid tissue isn't yet well developed. Children and adults also develop preauricular lymphadenopathy, and children may develop otitis media as a complication. Inclusion conjunctivitis may persist for weeks or months, possibly with superficial corneal involvement.

## Diagnosis

Clinical features and a history of sexual contact with an infected individual suggest inclusion conjunctivitis.



**CONFIRMING DIAGNOSIS** Examination of Giemsa-stained conjunctival scraping reveals cytoplasmic inclusion bodies in conjunctival epithelial cells and is effective in detecting chlamydial infection in infants. The direct fluorescent monoclonal antibody and enzyme-linked immunosorbent assay are most effective in adults.

## Treatment

Because infection isn't limited to the eye in neonates, infants, or adults, systemic antimicrobial treatment is necessary. In infants, effective therapy is achieved with erythromycin. Adults may be given tetracycline, doxycycline, or erythromycin.

Prophylactic tetracycline or erythromycin ointment is applied once, 1 hour after delivery. However, this treatment hasn't been found to be significantly more effective than Credé procedure (1% silver nitrate).

## Special Considerations

- ◆ Keep the patient's eyes as clean as possible, using sterile technique. Clean the eyes from the inner to the outer canthus. Apply warm soaks as needed. Record the amount and color of drainage.
- ◆ Remind the patient not to rub the eyes, which can irritate them.
- ◆ If the patient's eyes are sensitive to light, keep the room dark or suggest wearing dark glasses.



**PREVENTION** *Take these actions to prevent further spread of inclusion conjunctivitis:*

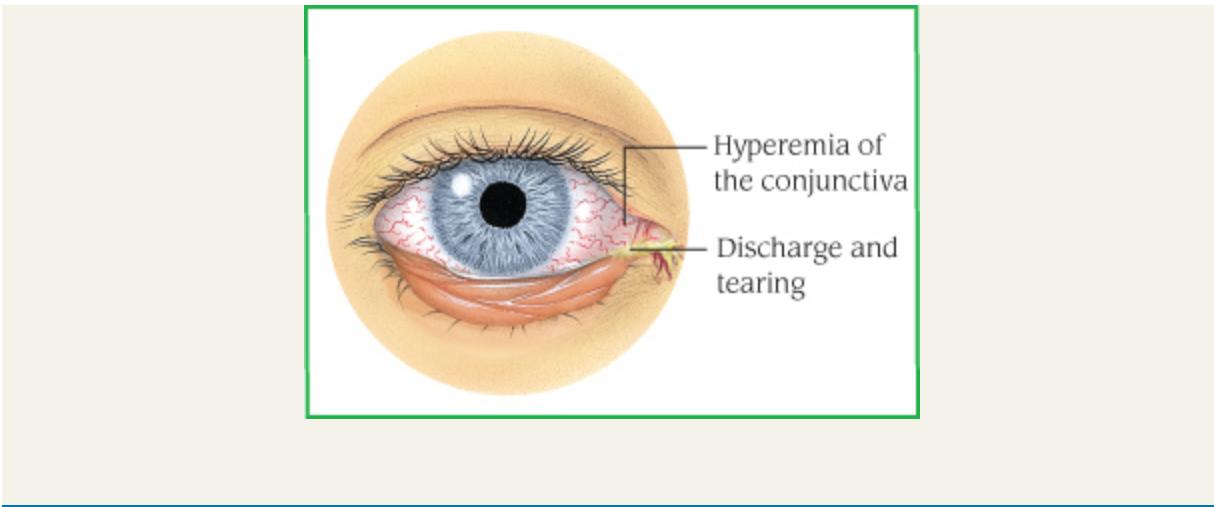
- ◆ *Wash hands thoroughly before and after administering eye medications.*
- ◆ *Suggest genital examination of the mother of an infected neonate or of any adult with inclusion conjunctivitis.*
- ◆ *Obtain a history of recent sexual contacts, so they can be examined for chlamydial infection.*

## CONJUNCTIVITIS

Conjunctivitis is characterized by hyperemia of the conjunctiva due to infection, allergy, or chemical reactions. (See *Recognizing conjunctivitis*.) This disorder usually occurs as benign, self-limiting pinkeye; it may also be chronic, possibly indicating degenerative changes or damage from repeated acute attacks.

### Recognizing Conjunctivitis

Itching is the hallmark of allergy. Giant papillae resembling cobblestones may be seen on the palpebral conjunctiva, as shown here.



## Pathophysiology

The most common causative organisms include:

- ◆ bacterial—*S. aureus*, *S. pneumoniae*, *Neisseria gonorrhoeae*, *Neisseria meningitidis*
- ◆ chlamydial—*C. trachomatis* (inclusion conjunctivitis)
- ◆ viral—adenovirus types 3, 7, and 8; herpes simplex virus, type 1

Other causes include allergic reactions to pollen, grass, topical medications, air pollutants, smoke, or unknown seasonal allergens (vernal conjunctivitis); environmental (wind, dust, and smoke) and occupational irritants (acids and alkalies); and a hypersensitivity to contact lenses or solutions.

Vernal conjunctivitis (so-called because symptoms tend to be worse in the spring) is a severe form of immunoglobulin E-mediated mast cell hypersensitivity reaction. This form of conjunctivitis is bilateral. It usually begins at age 3 to 5 years and persists for about 10 years. It's sometimes associated with other signs of allergy commonly related to pollens, asthma, and allergic rhinitis.

Epidemic keratoconjunctivitis is an acute, highly contagious viral conjunctivitis caused by adenovirus types 8 and 19. On occasion it is complicated by visual loss due to corneal subepithelial infiltrates. Healthcare providers must be careful to wash their hands and sterilize equipment to prevent the spread of this disease.

In the Western hemisphere, conjunctivitis is probably the most common eye disorder.

## Complications

- ◆ Corneal infiltrates
- ◆ Corneal ulcers
- ◆ Reinfection

## Signs and Symptoms

Conjunctivitis commonly produces hyperemia of the conjunctiva, sometimes accompanied by discharge, tearing and, with corneal involvement, pain and photophobia. It generally doesn't affect vision. Conjunctivitis usually begins in one eye and rapidly spreads to the other by contamination of towels, washcloths, or the patient's own hand.

Acute bacterial conjunctivitis (pinkeye) usually lasts only 2 weeks. The patient typically complains of itching, burning, and the sensation of a foreign body in the eye. The eyelids show a crust of sticky, mucopurulent discharge. If the disorder is due to *N. gonorrhoeae*, however, the patient exhibits a profuse, purulent discharge. In that case, treatment is required to avoid severe complications, including corneal perforation and endophthalmitis.

Viral conjunctivitis produces copious tearing with minimal exudate, and enlargement of the preauricular lymph node. Some viruses follow a chronic course; others last 2 to 3 weeks and are self-limiting.

## Diagnosis

Examination includes inspection of the eyelids, conjunctiva, and cornea. Regional lymph nodes should also be palpated. In children, possible systemic symptoms include sore throat or fever, if the conjunctivitis is suspected of being of adenoviral origin.

Lymphocytes are predominant in stained smears of conjunctival scrapings if conjunctivitis is caused by a virus. Polymorphonuclear cells (neutrophils) predominate if conjunctivitis is due to bacteria; eosinophils, if it's allergy-related. Culture and sensitivity tests identify the causative bacterial organism and indicate appropriate antibiotic therapy.

## Treatment

Treatment for conjunctivitis varies with the cause. Bacterial conjunctivitis requires topical application of the appropriate broad-spectrum antibiotic.

Although viral conjunctivitis resists treatment, a sulfonamide or broad-spectrum antibiotic eyedrops may prevent a secondary infection. Patients may be contagious for several weeks after onset. The most important aspect of treatment is preventing transmission. Herpes simplex infection generally responds to treatment with trifluridine drops or vidarabine ointment or oral acyclovir, but the infection may persist for 2 to 3 weeks. Treatment for vernal (allergic) conjunctivitis includes administration of corticosteroid drops followed by cromolyn sodium, cold compresses to relieve itching, and, occasionally, oral antihistamines.

Instillation of a one-time dose of erythromycin or 1% silver nitrate solution (Credé procedure) into the eyes of neonates prevents gonococcal conjunctivitis.

## Special Considerations

- ◆ Apply compresses and therapeutic ointment or drops, as ordered. Don't irrigate the eye, as this will spread the infection. Have the patient wash hands before using the medication. Instruct the patient to use clean washcloths or towels frequently to avoid infecting the other eye. (See *Preventing conjunctivitis*, page 606.)
- ◆ Teach the patient to instill eyedrops and ointments correctly—without touching the bottle tip to the eye or lashes.
- ◆ Remind the patient that the ointment will cause blurred vision.
- ◆ Stress the importance of safety glasses for the patient who works near chemical irritants.
- ◆ Notify public health authorities if cultures show *N. gonorrhoeae*.
- ◆ Ensure appropriate follow-up (e.g., patients on corticosteroid drops should have their IOP monitored periodically).



## PREVENTION PREVENTING CONJUNCTIVITIS

To prevent conjunctivitis from occurring or recurring, teach your patient to practice good hygiene. Encourage the following prevention tips.

### Practice Good Hygiene

To encourage good eye hygiene, teach proper hand-washing technique because bacterial and viral conjunctivitis are highly contagious. Stress the risk of spreading infection to family members by sharing washcloths, towels, and pillows. Suggest the use of tissues or disposable wipes to reduce the risk of transmission from contaminated linens. Caution the patient against rubbing the infected eye, which could spread infection to the other eye.

### **Use Cosmetics Carefully**

If the patient uses eye cosmetics, instruct the patient not to share them. Also, encourage the patient to replace eye cosmetics regularly.

### **Keep Contact Lenses Clean**

If the patient wears contact lenses, teach the patient to handle and clean contact lenses properly. Also, advise the patient to stop wearing contact lenses until the infection clears.

### **Avoid Contact With Contagious People**

Because conjunctivitis is highly contagious, particularly among children, infected children should avoid close contact with other children. Warn the patient with “cold sores” to avoid kissing others on the eyelids to prevent the spread of the disease.

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## **TRACHOMA**

The most common cause of preventable blindness in underdeveloped areas of the world, trachoma is a chronic form of keratoconjunctivitis. This infection is usually confined to the eye but may have a systemic component. Although trachoma itself is self-limiting, it causes permanent damage to the cornea and conjunctiva by scarring the lids, and it results in secondary infections that can lead to blindness. (See *What happens in trachoma*, page 607.) Early diagnosis and treatment (before trachoma results in scar formation) ensure recovery but without immunity to reinfection.



**CONFIRMING DIAGNOSIS** *Microscopic examination of a Giemsa-stained conjunctival scraping confirms the diagnosis by showing cytoplasmic*

*inclusion bodies, some polymorphonuclear reaction, plasma cells, and large macrophages containing phagocytosed debris.*

## **Pathophysiology**

Trachoma results from infection with *C. trachomatis*, a gram-negative obligate intracellular bacterium. These organisms are transmitted from eye to eye by flies and gnats and through hand-to-eye contact in endemic areas.

Trachoma is spread by close contact between family members or among schoolchildren. It's prevalent in Africa, Latin America, and Asia, particularly in children. Other predisposing factors include poverty and poor hygiene due to lack of water. Patients in hot, dusty climates are at greater risk.

## **Complications**

- ◆ Conjunctival and corneal scarring
- ◆ Deformities of the eyelid
- ◆ Vision loss

## **Signs and Symptoms**

Trachoma begins with a mild infection resembling bacterial conjunctivitis (visible conjunctival follicles, red and edematous eyelids, pain, photophobia, tearing, and exudation).

After about 1 month, if the infection is untreated, conjunctival follicles enlarge into inflamed papillae that later become yellow or gray. At this stage, small blood vessels invade the superior cornea under the upper lid.

Eventually, severe scarring and contraction of the eyelids cause entropion; the eyelids turn inward and the lashes rub against the cornea, producing corneal scarring and visual distortion. In late stages, severe conjunctival scarring may obstruct the lacrimal ducts and cause dry eyes.

## **Diagnosis**

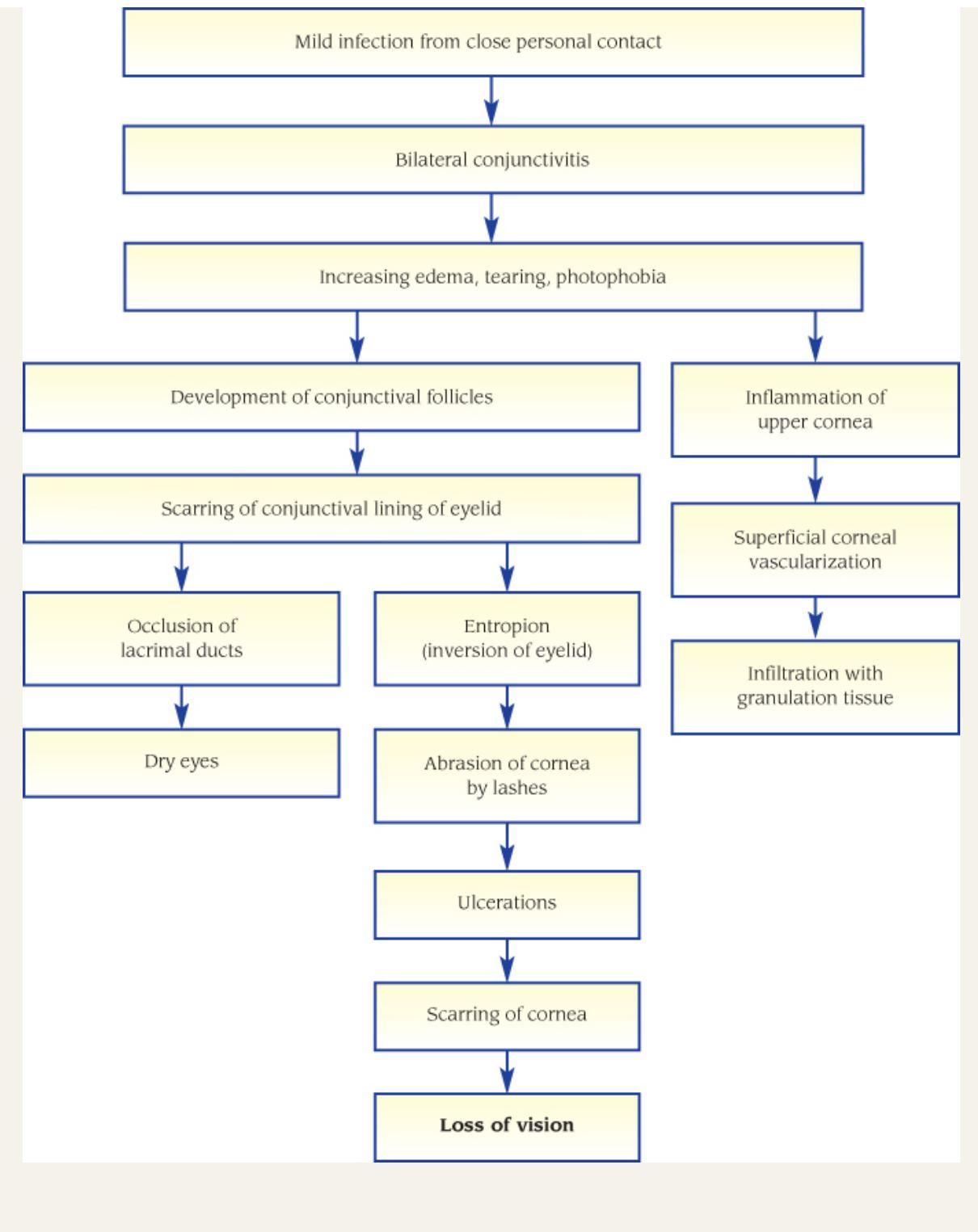
Follicular conjunctivitis with corneal infiltration, and upper lid or conjunctival scarring suggest trachoma, especially in endemic areas, when these symptoms persist longer than 3 weeks.

## PATHOPHYSIOLOGY



### WHAT HAPPENS IN TRACHOMA

Trachoma results from infection with *C. trachomatis* and in its early stages resembles bacterial conjunctivitis. If untreated, this chronic infection can spread to the cornea and lead to scarring and, eventually, to blindness.



## Treatment

Primary treatment for trachoma consists of topical or systemic antibiotic therapy with erythromycin (and its derivatives), doxycycline, or

sulfonamides. Severe entropion requires surgical correction.



**ALERT** *Tetracycline is contraindicated in pregnant women, because it may adversely affect the fetus, and in children younger than age 7, in whom it may permanently discolor teeth.*

Because trachoma is contagious and reinfection is common, some physicians suggest treating entire villages with high incidence rates with antibiotic therapy.

## Special Considerations

Patient teaching is essential:

- ◆ Emphasize the importance of hand washing and making the best use of available water supplies to maintain good personal hygiene.
- ◆ Because no definitive preventive measure exists (vaccines offer temporary and partial protection, at best), stress the need for strict compliance with the prescribed drug therapy.
- ◆ If ordered, teach the patient or family how to instill eyedrops correctly.
- ◆ Stress the importance of not sharing contaminated items, such as towels, handkerchiefs, and eye makeup.
- ◆ Emphasize the need for facial cleanliness.



**PREVENTION** *To prevent trachoma, warn the patient not to allow flies or gnats to settle around the eyes.*

## Corneal Disorders

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### KERATITIS

An inflammation of the cornea, keratitis may result from bacterial, fungal, or viral infection. If untreated, the infection can lead to blindness.

### Pathophysiology

The most common cause of keratitis is infection by herpes simplex virus, type 1 (causing a *dendritic corneal ulcer*, so named because of its characteristic branched lesion on the cornea resembling the veins of a leaf).

Bacterial corneal ulcers frequently occur as a result of an infected corneal abrasion or a contaminated contact lens. Fungal keratitis is more frequently encountered in tropical climates, after trauma, or in the elderly. Poor lid closure can result in exposure keratitis. Chemicals accidentally splashed into the eye and exposure to ultraviolet light (sunlamps, sunlight, or welding arcs) also can produce keratitis.



**ALERT** *An ocular chemical burn is an ophthalmic emergency. Patients need copious irrigation of the affected eye until the pH of the tears is back to physiologic levels.*

## Complications

- ◆ Irregular astigmatism
- ◆ Corneal scarring or perforation

## Signs and Symptoms

The patient presents with decreased vision, discomfort ranging from mild irritation to acute pain, tearing, and photophobia. On gross examination with a penlight, the corneal light reflex may appear distorted or dull. When keratitis results from exposure, it usually affects the lower portion of the cornea.

## Diagnosis



**CONFIRMING DIAGNOSIS** *Visual acuity may be decreased if the lesion is central. Slit-lamp examination confirms keratitis. Staining the eye with a sterile fluorescein strip enables the examiner to discern the extent and depth of any corneal lesion.*

Patient history may reveal a recent infection of the upper respiratory tract accompanied by cold sores, if the etiology is herpetic. It is important to take a contact lens history (e.g., overnight wear or improper cleaning).

## Treatment

Treatment for acute keratitis due to herpes simplex virus consists of trifluridine eyedrops, vidarabine ointment, and/or oral acyclovir. A broad-spectrum antibiotic may prevent secondary bacterial infection. Dendritic

keratitis may become chronic with recurrent episodes. Bacterial corneal ulcers require intense topical eyedrop instillation every half hour for the first 48 hours with two broad-spectrum antibiotics. Long-term topical therapy may be necessary. (Corticosteroid therapy is contraindicated in dendritic keratitis or any other viral or fungal disease of the cornea.) Fungal keratitis is treated with polyhexamethylene biguanide.

Exposure keratitis is treated with ointment at night and frequent instillation of artificial tears during the day. A plastic bubble shield may prevent tear evaporation. Vision may be restored by penetrating keratoplasty (corneal transplant) in blindness resulting from corneal scarring.

## Special Considerations

- ◆ Protect the exposed corneas of unconscious patients by cleaning the eyes daily, applying moisturizing ointment, or covering the eyes with an eye shield.
- ◆ Be aware that the patient with a red eye may have keratitis. Check for a history of contact lens wear, cold sores, or recent foreign body sensation. Refer the patient for slit-lamp examination as soon as possible for intense treatment.

## CORNEAL ABRASION

A corneal abrasion is a scratch on the surface epithelium of the cornea. With treatment, the prognosis is usually good.

### Pathophysiology

Eye trauma or a foreign body (such as a cinder or a piece of dust, dirt, or grit) on the cornea or under the lid are the most common causes of an abrasion.

A corneal scratch produced by a fingernail, a piece of paper, or other organic substance may cause a persistent lesion. The epithelium doesn't always heal properly, and a recurrent corneal erosion may develop, with delayed effects more severe than the original injury.

In the United States, corneal abrasions are a common ophthalmologic cause of emergency department visits. Incidence is highest among younger, physically active individuals; corneal abrasions are rare in elderly people.

## Complications

- ◆ Corneal erosion
- ◆ Corneal ulceration
- ◆ Permanent vision loss

## Signs and Symptoms

A corneal abrasion typically produces redness, increased tearing, discomfort with blinking, a sensation of “something in the eye” and, because the cornea is richly endowed with nerve endings from the trigeminal nerve (cranial nerve V), pain disproportionate to the size of the injury. It may also affect visual acuity, depending on the size and location of the injury.

## Diagnosis

History of eye trauma or prolonged wearing of contact lenses and typical symptoms suggest corneal abrasion.

 **CONFIRMING DIAGNOSIS** *Staining the cornea with fluorescein stain confirms the diagnosis: The injured area appears green when examined with a cobalt blue light. Slit-lamp examination discloses depth and allows measurement of the abrasion.*

Examining the eye with a flashlight may reveal a foreign body on the cornea; the eyelid must be everted to check for a foreign body embedded under the lid.

Before beginning treatment, a test to determine visual acuity provides a medical baseline and a legal safeguard.

## Treatment

Topical anesthetic eyedrops are instilled in the affected eye before removal of a superficial foreign body, using a foreign body spud. A rust ring on the cornea must be removed with an ophthalmic burr.

Treatment includes instillation of broad-spectrum antibiotic eyedrops in the affected eye every 3 to 4 hours. Application of a pressure patch prevents further corneal irritation when the patient blinks. If the patient wears contact lenses, advise the patient to abstain from wearing the lenses until the corneal abrasion heals.

## Special Considerations

- ◆ Assist with examination of the eye. Check visual acuity before beginning treatment.
- ◆ If a foreign body is visible, carefully irrigate with normal saline solution.
- ◆ Tell the patient with an eye patch to leave it in place as directed. Warn that a patch alters depth perception, so advise caution in daily activities, such as climbing stairs or stepping off a curb.
- ◆ Reassure the patient that the corneal epithelium usually heals in 24 to 48 hours.
- ◆ Stress the importance of instilling antibiotic eyedrops, as ordered, because an untreated corneal abrasion, if infected, can lead to a corneal ulcer and permanent vision loss. Teach the patient the proper way to instill eye medications.



**PREVENTION** Emphasize the importance of wearing safety glasses to protect a worker's eyes from flying fragments. Also review instructions for wearing and caring for contact lenses, to prevent further trauma. Encourage use of sunglasses.

## CORNEAL ULCERS

A major cause of blindness worldwide, ulcers produce corneal scarring or perforation. They occur in the central or marginal areas of the cornea, vary in shape and size, and may be singular or multiple. Marginal ulcers are the most common form. Prompt treatment (within hours of onset) can prevent visual impairment.

## Pathophysiology

Corneal ulcers generally result from bacterial, viral, fungal, or protozoan infections. Common bacterial sources include *S. aureus*, *Pseudomonas aeruginosa*, *Streptococcus viridans*, *S. (Diplococcus) pneumoniae*, and *Moraxella liquefaciens*; viral sources comprise herpes simplex type 1, variola, vaccinia, and varicella-zoster viruses; and common fungal sources are *Candida*, *Fusarium*, and *Cephalosporium*.

Other causes include trauma, exposure, reactions to bacterial infections, toxins, trichiasis, entropion, allergens, and wearing of contact lenses. (See *What happens in corneal ulceration*.) Tuberculoprotein causes a classic

phlyctenular keratoconjunctivitis, vitamin A deficiency results in xerophthalmia, and fifth cranial nerve lesions lead to neurotropic ulcers.

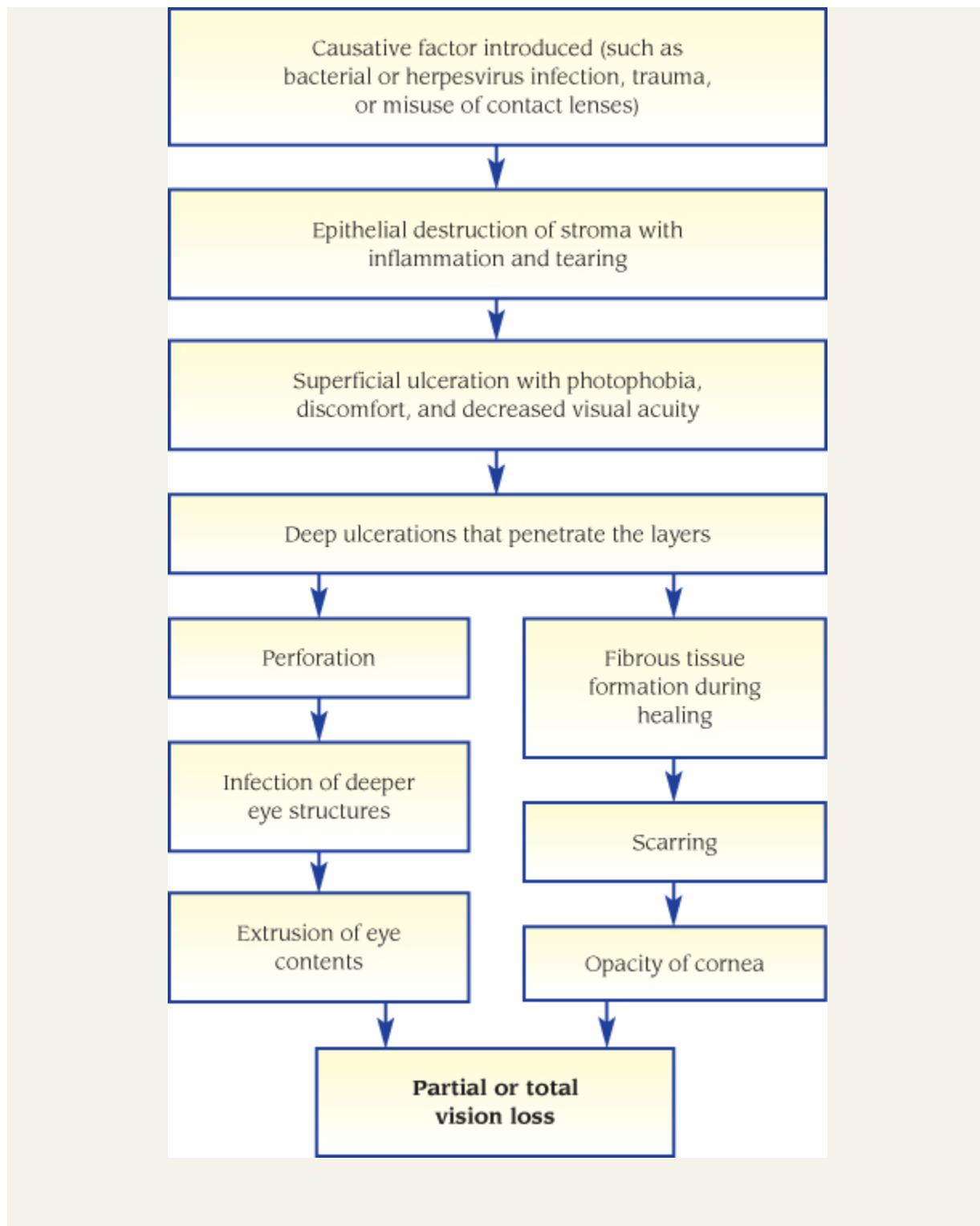
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## **PATHOPHYSIOLOGY**

### **WHAT HAPPENS IN CORNEAL ULCERATION**

Corneal ulcers can be caused by infection (protozoan, bacterial, viral, or fungal), trauma, exposure, toxins, contact lenses, or allergens. Scarring or perforation can cause changes in the eye structure and can lead to partial or total vision loss.



## Complications

- ◆ Corneal scarring
- ◆ Loss of the eye

- ◆ Vision loss
- ◆ Irregular astigmatism
- ◆ Corneal perforation

## Signs and Symptoms

Typically, corneal ulceration begins with pain and foreign body sensation (aggravated by blinking) and photophobia, followed by increased tearing. The eye may appear injected. If a bacterial ulcer is present, purulent discharge is possible.

## Diagnosis

A history of trauma or use of contact lenses and flashlight examination that reveals irregular corneal surface suggest corneal ulcer. Exudate may be present on the cornea, and a hypopyon (accumulation of white cells in the anterior chamber) may appear as a white crescent moon inside the eye that moves when the head is tilted.



**CONFIRMING DIAGNOSIS** *Fluorescein dye, instilled in the conjunctival sac, stains the outline of the ulcer and confirms the diagnosis.*

Culture and sensitivity testing of corneal scrapings may identify the causative bacteria or fungus, and may indicate appropriate antibiotic or antifungal therapy.

## Treatment

Prompt treatment is essential for all forms of corneal ulcer to prevent complications and permanent visual impairment. Treatment usually consists of topical broad-spectrum antibiotics until culture results identify the causative organism. The goals of treatment are to eliminate the underlying cause of the ulcer and to relieve pain:

- ◆ Fungi—topical instillation of polyhexamethylene biguanide for *Fusarium*, *Cephalosporium*, and *Candida*.
- ◆ Herpes simplex virus type 1—topical application of trifluridine drops or vidarabine ointment. Corneal ulcers resulting from a viral infection often recur, requiring further treatment with trifluridine.
- ◆ Hypovitaminosis A—correction of dietary deficiency or gastrointestinal (GI) malabsorption of vitamin A.

- ◆ Infection by *P. aeruginosa*—fluoroquinolones, administered topically and by subconjunctival injection, tobramycin I.V. Because this type of corneal ulcer spreads so rapidly, it can cause corneal perforation and loss of the eye within 48 hours. Immediate treatment and isolation of hospitalized patients are required.
- ◆ Neurotropic ulcers or exposure keratitis—frequent instillation of artificial tears or lubricating ointments and use of a plastic bubble eye shield.
- ◆ Varicella-zoster virus—topical erythromycin ointment applied three to four times daily to prevent secondary infection. These lesions are unilateral, following the pathway of the fifth cranial nerve, and are typically quite painful. Give analgesics and oral acyclovir as ordered. Associated anterior uveitis requires cycloplegic eyedrops. Watch for signs of secondary glaucoma (transient vision loss and halos around lights).



**ALERT** *Treatment for a corneal ulcer due to bacterial infection should never include an eye patch because patching creates the dark, warm, moist environment ideal for bacterial growth.*

## Special Considerations

- ◆ Keep the room darkened and orient the patient as necessary.
- ◆ Teach the patient how to properly clean and wear contact lenses to prevent a recurrence.



**PREVENTION** *Encourage your patient to:*

- ◆ *seek treatment early for eye infections*
- ◆ *wash hands before handling contact lenses*
- ◆ *avoid wearing contact lenses overnight*

## Uveal Tract, Retinal, and Lens Disorders

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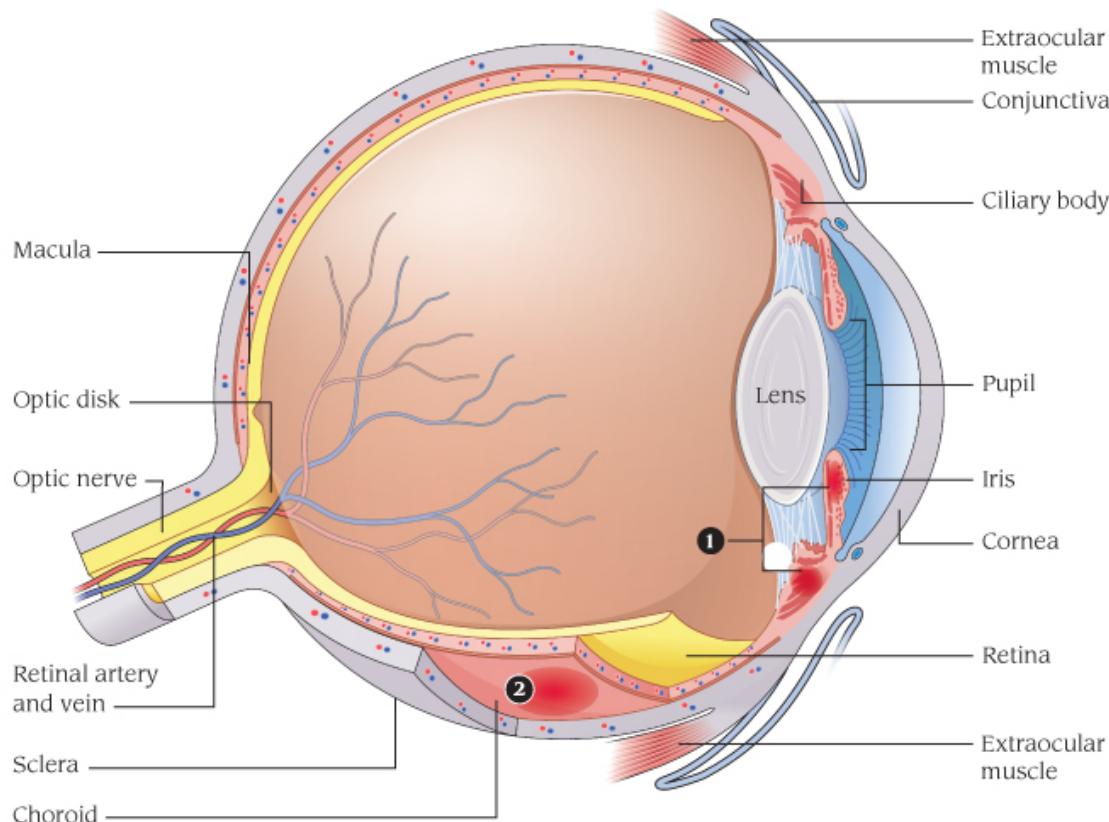
### UVEITIS

Uveitis is inflammation of the uveal tract. It occurs as anterior uveitis, which affects the iris (iritis) or both the iris and the ciliary body (iridocyclitis); as posterior uveitis, which affects the choroid (choroiditis) or both the choroid and the retina (chorioretinitis); or as panuveitis, which affects the entire uveal tract. Although clinical distinction isn't always possible, anterior uveitis occurs in two forms—granulomatous and nongranulomatous. (See *The uveal tract and causes of uveitis*, page 612.)



## **PATHOPHYSIOLOGY THE UVEAL TRACT AND CAUSES OF UVEITIS**

Uveitis is the inflammation of the uveal tract. It occurs as anterior uveitis, posterior uveitis, or panuveitis (affects the entire uveal tract).



**① Anterior uveitis (iritis and iridocyclitis)**

Acute

- Idiopathic
- Trauma/surgery
- Virus or *Chlamydia* infection

Chronic

- Juvenile rheumatoid arthritis and related autoimmune conditions
- Sarcoidosis
- Herpesvirus infection
- Syphilis
- Sympathetic (autoimmune) ophthalmritis

**② Posterior uveitis (choroiditis)**

Any of ① plus:

- Diabetic retinopathy
- Opportunistic infections (toxoplasmosis, cytomegalovirus) in patients with AIDS or other immune deficiency

Granulomatous uveitis was once thought to be caused by tuberculosis bacilli; nongranulomatous uveitis, by streptococci. Although this isn't true, the terms are still used. Untreated anterior uveitis may result in elevated IOP, leading to vision loss. With immediate treatment, anterior uveitis usually subsides after a few days to several weeks; however, recurrence can occur. Posterior uveitis may lead to vision loss if the macula is involved.

## Pathophysiology

Typically, uveitis is idiopathic. However, it can result from allergy, bacteria, viruses, fungi, chemicals, trauma, or surgery; or it may be associated with systemic diseases, such as rheumatoid arthritis, ankylosing spondylitis, and toxoplasmosis.

Uveitis occurs in 15 of every 100,000 people.

## Complications

- ◆ Cataracts
- ◆ Glaucoma
- ◆ Retinal detachment
- ◆ Vision loss

## Signs and Symptoms

Anterior uveitis produces moderate to severe unilateral eye pain; severe ciliary injection; photophobia; tearing; a small, nonreactive pupil; and blurred vision (due to the increased number of cells in the aqueous humor). It sometimes produces deposits called *keratic precipitates* on the back of the cornea. The iris may adhere to the lens, causing posterior synechiae and pupillary distortion. Onset may be acute or insidious.

Posterior uveitis begins insidiously, with complaints of slightly decreased or blurred vision or floating spots. Posterior uveitis may be acute or chronic, and it may affect one or both eyes. Retinal damage caused by lesions from toxoplasmosis and retinal detachments may occur. Refer the patient to an eye care practitioner for dilated fundus examination and treatment for systemic diseases.

## Diagnosis

 **CONFIRMING DIAGNOSIS** In anterior and posterior uveitis, a slit-lamp examination shows a “flare and cell” pattern, which looks like particles dancing in a sunbeam. With a special lens, slit-lamp and ophthalmoscopic examination can also identify active inflammatory fundus lesions involving the retina and choroid, although a hazy vitreous may obscure the view.

In posterior uveitis, serologic tests may be used to rule out toxoplasmosis, other infections, or inflammatory etiologies.

## Treatment

Uveitis requires vigorous and prompt management, which includes treatment for any known underlying cause—corticosteroids with antibiotic therapy for infectious diseases and immune suppression therapy for autoimmune diseases—and application of a topical cycloplegic, such as 1% atropine sulfate, and of topical corticosteroids applied three to four times daily. For severe uveitis, therapy includes oral systemic corticosteroids.



**ALERT** Long-term steroid therapy can cause a rise in IOP and cataracts. Carefully monitor IOP during acute inflammation. If IOP rises, therapy should include an antiglaucoma medication, such as brimonidine, an alpha<sub>2</sub>-adrenergic agonist, dorzolamide, a sulfonamide, or timolol (a beta-adrenergic blocker).

Occasionally, posterior uveitis requires systemic immunosuppression.

## Special Considerations

- ◆ Encourage rest during the acute phase.
- ◆ Teach the patient the proper method of instilling eyedrops.
- ◆ Suggest the use of dark glasses to ease the discomfort of photophobia.
- ◆ Instruct the patient to watch for and report adverse effects of systemic corticosteroid therapy (e.g., edema or muscle weakness).
- ◆ Stress the importance of follow-up care for IOP checks while the patient is taking steroids. Tell the patient to seek treatment immediately at the first sign of iritis.

## RETINAL DETACHMENT

Retinal detachment occurs when the outer retinal pigment epithelium splits from the neural retina, creating subretinal space. This space then fills with fluid, called *subretinal fluid*. Retinal detachment usually involves only one eye, but may later involve the other eye. Surgical reattachment is usually successful. However, the prognosis for good vision depends on which area of the retina has been affected.

## Pathophysiology

Any retinal tear or hole allows the liquid vitreous to seep between the retinal layers, separating the retina from its choroidal blood supply. Predisposing factors include myopia, intraocular surgery, and trauma. In adults, retinal detachment usually results from degenerative changes of aging, which cause a spontaneous retinal hole. Perhaps the influence of trauma explains why retinal detachment is twice as common in males. Retinal detachment may also result from seepage of fluid into the subretinal space (because of inflammation, tumors, or systemic diseases) or from traction that's placed on the retina by vitreous bands or membranes (due to proliferative diabetic retinopathy, posterior uveitis, or a traumatic intraocular foreign body).

Retinal detachment is rare in children, but occasionally can develop as a result of retinopathy of prematurity, tumors (retinoblastomas), trauma, or myopia (which tends to run in families).

In the United States, about 10,000 people per year are affected by retinal detachments.

## Complications

- ◆ Severe vision impairment
- ◆ Blindness

## Signs and Symptoms

Initially, the patient may complain of floating spots and recurrent flashes of light (photopsia). However, as detachment progresses, gradual, painless vision loss may be described as a veil, curtain, or cobweb that obscures a portion of the visual field.

## Diagnosis

 **CONFIRMING DIAGNOSIS** *Diagnosis depends on ophthalmoscopy after full pupil dilation. Examination shows the usually transparent retina as gray and opaque; in severe detachment, it reveals folds in the retina and ballooning out of the area. Indirect ophthalmoscopy is used to search for retinal tears. Ultrasound is performed if the lens is opaque.*

## Treatment

Treatment depends on the location and severity of the detachment. It may include restriction of eye movements and complete bed rest until surgical reattachment is done. A hole in the peripheral retina can be treated with cryotherapy; in the posterior portion, with laser therapy. Retinal detachment usually requires a scleral buckling procedure or a vitrectomy to reattach the retina. Basic salt solution is used to reposition the retina while the vitreous is removed.

Certain types of uncomplicated retinal detachment may be treated by pneumatic retinopexy, in which an expansile gas is initially injected into the vitreous cavity and the patient's head is positioned to facilitate retina reattachment. This procedure can be performed under local anesthesia.

## **Special Considerations**

- ◆ Provide emotional support because the patient may be understandably distraught about loss of vision.
- ◆ During transportation, position the patient's head so that the detached portion of the retina will fall back with the aid of gravity.
- ◆ To prepare for surgery, wash the patient's face with no-tears shampoo. Give antibiotics and cycloplegic-mydratic eyedrops.
- ◆ Postoperatively, position the patient as recommended by the surgeon. Discourage straining at stool, bending down, hard coughing, sneezing, or vomiting, which can raise IOP. Antiemetics may be indicated.
- ◆ Protect the patient's eye with a shield or glasses.
- ◆ To reduce edema and discomfort, apply ice packs as ordered. Administer pain medication, as ordered, for eye pain.
- ◆ After removing the eye shield, gently clean the eye and administer steroid-antibiotic eyedrops, as ordered. Use cold compresses to decrease swelling and pain.
- ◆ Administer analgesics as needed, and report persistent pain. Teach the patient how to properly instill eyedrops, and emphasize compliance and follow-up care. Suggest dark glasses to compensate for light sensitivity caused by cycloplegia.

## **VASCULAR RETINOPATHIES**

Vascular retinopathies are noninflammatory retinal disorders that result from interference with the blood supply to the eyes. The five most common types of vascular retinopathy are central retinal artery occlusion, central

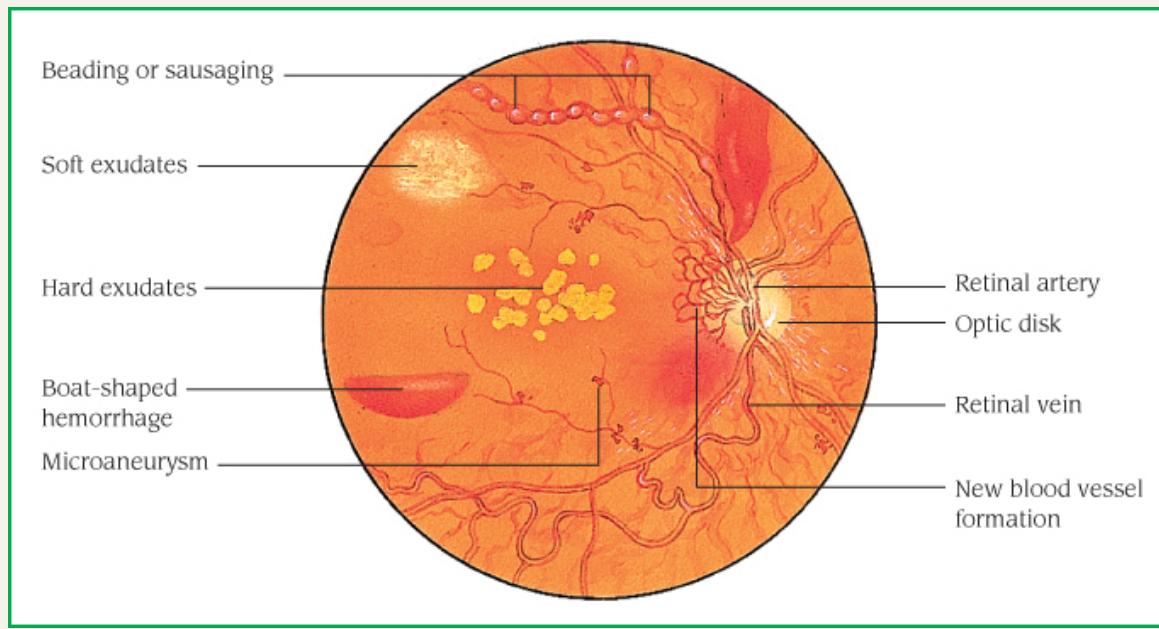
retinal vein occlusion, diabetic retinopathy, hypertensive retinopathy, and sickle cell retinopathy.

## Pathophysiology

When one of the arteries maintaining blood circulation in the retina becomes obstructed, the diminished blood flow causes visual deficits. (See *Anatomy of vascular retinopathy*.)

## Anatomy Of Vascular Retinopathy

Vascular changes that occur with retinopathy, as seen by an ophthalmoscope, are depicted below.



Central retinal artery occlusion may be idiopathic or may result from embolism, atherosclerosis, infection, or conditions that retard blood flow, such as temporal arteritis, carotid occlusion, and heart failure. This occlusion is rare, occurs unilaterally, and usually affects elderly patients. However, if it occurs in a younger person, the obstruction may have originated in the heart (such as embolization from plaque material from valve vegetations) and should be investigated accordingly.

Causes of central retinal vein occlusion include atherosclerosis, hypertension, optic disk edema, hypercoagulable states (polycythemia, leukemia, or sickle cell disease), glaucoma, retrobulbar compression (such as an orbital tumor), and drugs such as hormonal contraceptives. This form of vascular retinopathy is most prevalent in elderly patients and is characterized by impaired venous outflow.

Diabetic retinopathy results from type 1 or type 2 diabetes. Microcirculatory changes occur more rapidly when diabetes is poorly controlled. About 90% of patients with type 1 diabetes develop retinopathy within 20 years of onset of diabetes. In adults with diabetes, incidence increases with the duration of diabetes; 80% of patients who have had diabetes for 20 to 25 years develop retinopathy. This condition is a leading cause of acquired adult blindness.

Hypertensive retinopathy results from prolonged hypertensive disease, producing retinal vasospasm, and consequent damage and arteriolar narrowing.

Sickle cell retinopathy results from impaired ability of the sickled cell to pass through microvasculature, producing vaso-occlusion. This leads to microaneurysms, chorioretinal infarction, and retinal detachment.

## Complications

- ◆ Blindness
- ◆ Secondary glaucoma

## Signs and Symptoms

Central retinal artery occlusion produces sudden, painless, unilateral loss of vision (partial or complete). It may follow amaurosis fugax or transient episodes of unilateral loss of vision lasting from a few seconds to minutes, probably due to vasospasm. This condition typically causes permanent blindness. However, some patients experience spontaneous resolution within hours and regain partial vision.

Central retinal vein occlusion causes reduced visual acuity, allowing perception of only hand movement and light. This condition is painless, except when it results in secondary neovascular glaucoma (uncontrolled proliferation of weak blood vessels). The prognosis is poor—some patients with this condition develop secondary glaucoma within 3 to 4 months after occlusion.

Nonproliferative diabetic retinopathy produces changes in the lining of the retinal blood vessels that cause the vessels to leak plasma or fatty substances, which decrease or block blood flow (nonperfusion) within the retina. This disorder may also produce microaneurysms and small hemorrhages. Nonproliferative retinopathy causes no symptoms in some patients; in others, leakage of fluid into the macular region causes significant loss of central visual acuity (necessary for reading and driving) and diminished night vision.

Proliferative diabetic retinopathy produces fragile new blood vessels on the disk (neovascularization) and elsewhere in the fundus. These vessels can grow into the vitreous and then rupture, causing vitreous hemorrhage with corresponding sudden vision loss. Scar tissue that may form along the new blood vessels can pull on the retina, causing it to tear or even detach.

Symptoms of hypertensive retinopathy include blurred vision, often accompanied by headache. Ophthalmoscopic examination may reveal diffuse arteriolar narrowing, venular tortuosity, silver wire reflexes, macular stars, and swelling of the head of the optic nerve (disk edema). Severe, prolonged disease eventually produces blindness; mild, prolonged disease produces visual defects.

Symptoms of sickle cell retinopathy include peripheral arteriolar occlusions, peripheral arteriovenous anastomoses, sea fan neurovascular fronds, vitreous hemorrhage as tractional forces and vitreous collapse tear fragile neovascular membranes and, with advanced disease, severe vitreous traction and retinal detachment.

## Diagnosis

Check visual acuity and then vital signs, including blood pressure. Diagnosis is made on fundal examination with an ophthalmoscope. Determine if female patients are pregnant; hypertensive retinopathy may be an early sign of preeclampsia. (See *Diagnostic tests for vascular retinopathies*, page 616.)

## Treatment

No treatment has been shown to improve or resolve central retinal artery occlusion. However, an attempt is made to release the occlusion into the peripheral circulation. To reduce IOP, therapy includes acetazolamide I.V., eyeball massage, thrombolysis by intra-arterial injection or I.V., high

concentrations of inhaled oxygen, and anterior chamber paracentesis (to try to move the arterial obstruction into the peripheral field).

Therapy for central retinal vein occlusion may include aspirin, which acts as a mild anticoagulant. Patients with central retinal vein occlusion have reported improved vision after direct injection of tissue plasminogen activator into the retinal venous system. Laser photocoagulation can reduce the risk of neovascular glaucoma for some patients whose eyes have widespread capillary nonperfusion.

Treatment for nonproliferative diabetic retinopathy is prophylactic. Careful control of blood glucose levels reduces the severity of the retinopathy or may delay its onset. Patients with early symptoms of microaneurysms should have frequent eye examinations; children with diabetes should have an annual eye examination.

Treatment for proliferative diabetic retinopathy or severe macular edema is laser photocoagulation, which cauterizes the leaking blood vessels. Laser treatment may be focal (aimed at new blood vessels) or panretinal (placing burns throughout the peripheral retina). Despite treatment, neovascularization continues to proliferate, and vitreous hemorrhage, with or without retinal detachment, may follow. If the blood isn't absorbed in 6 weeks to 3 months, vitrectomy may restore partial vision. New agents, such as vascular endothelial growth factor inhibitors, can cause temporary regression of the abnormal blood vessels. This would be followed by panretinal laser treatment to prevent recurrence.

## Diagnostic Tests For Vascular Retinopathies

### Central Retinal Artery Occlusion

- ◆ Ophthalmoscopy (direct or indirect): shows blockage of retinal arterioles during a transient attack.
- ◆ Retinal examination: within 2 hours of onset, shows clumps or segmentation in the artery; later, milky white retina around the disk due to swelling and necrosis of ganglion cells caused by reduced blood supply; also shows a cherry-red spot in the macula that subsides after several weeks.
- ◆ Color Doppler tests: evaluate carotid occlusion with no need for arteriography.

- ◆ Physical examination: reveals the underlying cause of vascular retinopathy, for example, diabetes or hypertension.

### **Central Retinal Vein Occlusion**

- ◆ Ophthalmoscopy (direct or indirect): shows flame-shaped hemorrhages, retinal vein engorgement, white patches among hemorrhages, and edema around the disk.
- ◆ Color Doppler tests: confirm or rule out occlusion of blood vessels.
- ◆ Physical examination: reveals the underlying cause.

### **Diabetic Retinopathy**

- ◆ Ophthalmoscopic examination: shows retinal changes such as microaneurysms (earliest change), retinal hemorrhages and edema, venous dilation and beading, lipid exudates, fibrous bands in the vitreous, and growth of new blood vessels. Infarcts of the nerve fiber layer are observed.
- ◆ Fluorescein angiography: shows leakage of fluorescein from weak-walled vessels and “lights up” microaneurysms, differentiating them from true hemorrhages.
- ◆ History: of diabetes.

### **Hypertensive Retinopathy**

- ◆ Ophthalmoscopy (direct or indirect): in early stages, shows hard, shiny deposits; flame-shaped hemorrhages; silver wire appearance of narrowed arterioles; and nicking of veins where arteries cross them (atrioventricular nicking). In late stages, shows cotton wool patches, lipid exudates, retinal edema, papilledema due to ischemia and capillary insufficiency, hemorrhages, and microaneurysms in both eyes.
- ◆ Physical examination: reveals elevated blood pressure.
- ◆ History: of decreased vision, headache, and nausea.
- ◆ History: of hypertension, usually acute or malignant.

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Treatment for hypertensive retinopathy includes control of blood pressure with appropriate drugs, diet, and exercise. Treating the systemic

hypertension should improve the condition of the eyes. If left untreated, hypertensive retinopathy results in severe vision loss.

The treatment goal of sickle cell retinopathy is to reduce the risk of, or prevent or eliminate, retinal neovascularization. Patients with symptoms should receive follow-up care twice a year with ocular examinations and dilated retinal evaluation. Proliferative disease should be evaluated with fluorescein angiography and treated with panretinal photocoagulation. Cryotherapy hasn't been proven to be effective and has a high complication rate.

## Special Considerations

- ◆ Monitor a patient's blood pressure if there are complaints of occipital headache and blurred vision.
- ◆ Encourage a diabetic patient to comply with the prescribed regimen.
- ◆ For a patient with hypertensive retinopathy, stress the importance of complying with antihypertensive therapy.



**ALERT** *Arrange for immediate evaluation when a patient complains of sudden, unilateral loss of vision. Blindness may be permanent if treatment is delayed.*

## AGE-RELATED MACULAR DEGENERATION

Macular degeneration is the atrophy or degeneration of the macular region of the retina. Two types of age-related macular degeneration occur. The dry, or atrophic, form (which is the most common) is characterized by atrophic pigment epithelial changes and is most often associated with a slow, progressive, and mild vision loss. The wet, exudative form causes progressive visual distortion leading to vision loss. It's characterized by subretinal neovascularization that causes leakage, hemorrhage, and fibrovascular scar formation, which produce significant loss of central vision.

## Pathophysiology

Age-related macular degeneration results from underlying pathologic changes that occur primarily at the level of the retinal pigment epithelium and the adjacent structures (Bruch membrane and the choriocapillaris) in the macular region. Drusen, which are common in elderly people, appear as

yellow deposits beneath the pigment epithelium and may be prominent in the macula. No predisposing conditions have been identified; however, some forms of the disorder are hereditary.

Macular degeneration is the most common cause of legal blindness in adults, accounting for about 12% of blindness cases in the United States and for about 17% of new blindness cases. It's also one of the causes of severe irreversible loss of central vision in elderly people—by age 75, almost 15% of people have this condition. Whites have the highest incidence. Other risk factors are family history and cigarette smoking.

## Complications

- ◆ Blindness
- ◆ Metamorphopsia

## Signs and Symptoms

The patient notices a change in central vision. Initially, straight lines (e.g., of buildings) become distorted; later, a blank area appears in the center of a printed page (central scotoma).

## Diagnosis

- ◆ Ophthalmoscopy—fundus examination through a dilated pupil may reveal gross macular changes.
- ◆ I.V. fluorescein angiography—sequential photographs may show leaking vessels as fluorescein dye flows into the tissues from the subretinal neovascular net.
- ◆ Amsler grid—used to monitor metamorphopsia and the appearance of new scotomas.

## Treatment

Recent studies have shown that the dry form of age-related macular degeneration can be prevented and treated with daily vitamin therapy that includes lutein and zeaxanthin.

Laser photocoagulation reduces the incidence of severe vision loss in the patient with noncentral subretinal neovascularization, halting its progression. Injection of an antivascular endothelial growth factor agent into the vitreous is the most common treatment for serous (wet) age-related

macular degeneration. Timely treatment (before the formation of fibrovascular scars and retinal atrophy) can result in vision recovery. Recurrence may occur. Injections are repeated periodically, depending on the retinal evaluation.

## Special Considerations

- ◆ Inform the patient with bilateral central vision loss of the visual rehabilitation services available.
- ◆ Special devices, such as low-vision optical aids, are available to improve the quality of life in the patient with good peripheral vision.



**PREVENTION** Encourage early detection through regular eye examinations.

## CATARACT

The most common cause of correctable vision loss, a cataract is a gradually developing opacity of the lens or lens capsule of the eye. Cataracts commonly occur bilaterally, with each progressing independently. Exceptions are traumatic cataracts, which are usually unilateral, and congenital cataracts, which may remain stationary. The prognosis is generally good; surgery improves vision in 95% of affected people.

## Pathophysiology

Cataracts have various causes:

- ◆ Senile cataracts develop in elderly patients, probably because of degenerative changes in the chemical state of lens proteins.
- ◆ Congenital cataracts occur in neonates as genetic defects or as a sequela of maternal infections during the first trimester. Some cataracts are inherited in an autosomal dominant pattern.
- ◆ Traumatic cataracts develop after a foreign body injures the lens with sufficient force to allow aqueous or vitreous humor to enter the lens capsule. Trauma may also dislocate the lens.
- ◆ Complicated cataracts develop as secondary effects in patients with uveitis, glaucoma, or retinitis pigmentosa, or in the course of a systemic disease, such as diabetes, hypoparathyroidism, or atopic dermatitis. They can also result from exposure to ionizing radiation or infrared rays.

- ◆ Toxic cataracts result from drug or chemical toxicity with prednisone, ergot alkaloids, dinitrophenol, naphthalene, phenothiazines, or pilocarpine or from extended exposure to ultraviolet rays.

Cataracts occur as part of the aging process and are most prevalent in people older than age 70.

## Complication

- ◆ Vision loss

## Signs and Symptoms

Characteristically, a patient with a cataract experiences painless, gradual blurring and loss of vision. As the cataract progresses, the normally black pupil appears hazy, and when a mature cataract develops, the white lens may be seen through the pupil. Some patients complain of blinding glare from headlights when they drive at night; others complain of poor reading vision, and of an unpleasant glare and poor vision in bright sunlight. Patients with central opacities report better vision in dim light than in bright light because the cataract is nuclear and, as the pupils dilate, patients can see around the lens opacity.

## Diagnosis

On examination, visual acuity is decreased.



**CONFIRMING DIAGNOSIS** *Ophthalmoscopy or slit-lamp examination confirms the diagnosis by revealing a dark area in the normally homogeneous red reflex.*

## Treatment

Treatment consists of surgical extraction of the cataractous lens opacity and insertion of an intraocular artificial lens. Surgery is a same-day procedure. Surgical procedures include the following:

- ◆ Extracapsular cataract extraction (ECCE) removes the anterior lens capsule and cortex, leaving the posterior capsule intact. With this procedure, a posterior chamber intraocular lens (IOL) is implanted where the patient's own lens used to be. (A posterior chamber IOL is currently

the most common type used in the United States.) This procedure is appropriate for use in patients of all ages.

- ◆ Phacoemulsification uses ultrasonic vibrations to fragment and then emulsify the lens, which is then aspirated through a small incision.
- ◆ Intracapsular cataract extraction removes the entire lens within the intact capsule. This procedure is seldom performed today. ECCE with phacoemulsification has replaced it as the most commonly performed procedure.
- ◆ Discussion and aspiration can still be used for children with soft cataracts, but this procedure has largely been replaced by phacoemulsification.

Infection is the most serious complication of intraocular surgery. Wound dehiscence can occur but is seldom a complication because of the small incision and minute sutures that are used. Hyphema, pupillary block glaucoma, and retinal detachment still occasionally occur.

The patient with an IOL implant may experience improved vision shortly after surgery if there's no corneal or retinal pathology. Most IOLs correct for distance vision, but new IOLs are multifocal. However, the majority of patients will need either corrective reading glasses or a corrective contact lens, which will be fitted sometime between 4 and 6 weeks after surgery.

Where no IOL has been implanted, the patient may be given temporary aphakic cataract glasses; in about 4 to 8 weeks, he'll be refracted for glasses.

Some patients who have an ECCE develop a secondary membrane in the posterior lens capsule (which has been left intact), which causes decreased visual acuity. This membrane can be removed by the Nd:YAG laser, which cuts an area out of the center of the membrane, thereby restoring vision. Laser therapy isn't used to remove a cataract.

Posterior capsular opacification occurs in approximately 15% to 20% of all patients within 2 years after cataract surgery.

## Special Considerations

After surgery to extract a cataract:

- ◆ Because the patient will be discharged after recovering from anesthesia, remind the patient to return for a checkup the next day, and advise avoidance of activities that increase IOP such as straining.

- ◆ Urge the patient to protect the eye from accidental injury at night by wearing a plastic or metal shield with perforations; a shield or glasses should be worn for protection during the day.
- ◆ Before discharge, teach the patient to administer antibiotic ointment or drops to prevent infection and steroids to reduce inflammation; combination steroid-antibiotic eyedrops can also be used.
- ◆ Advise the patient to watch for the development of complications, such as a pain in the eye uncontrolled by analgesics, eye or eyelid redness, or decreased vision, and to report them immediately. These symptoms may indicate an infection.
- ◆ Caution the patient about activity restrictions, and advise that it will take several weeks to receive corrective reading glasses or lenses.



### **PREVENTION** Encourage your patient to:

- ◆ *quit smoking*
- ◆ *wear sunglasses*

## **RETINITIS PIGMENTOSA**

Retinitis pigmentosa is a group of hereditary disorders whose common feature is a gradual deterioration of the light-sensitive cells of the retina. Postmortem examination of the eyes reveals pigment cells that have clumped together as a result of the pigment epithelium budding off and settling within the layers of the retina. Retinitis pigmentosa often accompanies other hereditary disorders in several distinct syndromes—including Usher syndrome, in which sight and hearing are both affected; and Laurence–Moon–Biedl syndrome (most common), which is typified by visual destruction from retinitis pigmentosa, with obesity, mental retardation, polydactyly, hypogenitalism, and spastic paraplegia.

## **Pathophysiology**

Retinitis pigmentosa can be classified according to its inheritance pattern: autosomal dominant, autosomal recessive, and X-linked. Typically, in all forms of retinitis pigmentosa, the retinal rods slowly deteriorate. Clumps of pigment resembling bone corpuscles aggregate in the peripheral region of the retina and later involve the macular areas. Visual symptoms usually

appear between ages 10 and 30, though some children may become blind within the first year of life.

Retinitis pigmentosa affects 1 of every 4,000 people in the United States.

## Signs and Symptoms

Typically, night blindness occurs during the teen years. As the disease progresses, the visual field gradually constricts, causing tunnel or “gun-barrel” vision. Many people retain this tunnel of useful vision until quite late in life. The speed of vision loss varies considerably from person to person. However, blindness follows invasion of the macular region.

## Diagnosis

A detailed family history may imply predisposition to retinitis pigmentosa. In the patient whose history suggests this condition, the following tests help confirm diagnosis.

- ◆ Electroretinography shows a slower than normal or absent retinal response time.
- ◆ Fluorescein angiography visualizes white dots (areas of depigmentation) in the retina.
- ◆ Ophthalmoscopy may initially show normal fundi but later shows black pigmentary disturbance and white dots (depigmentation) in the retina.
- ◆ Visual field testing detects ring scotomata.

## Treatment

No cure exists for retinitis pigmentosa. However, vitamin A and E supplementation may slow degeneration. Researchers are working on the potential for tissue transplant, but research is still in its infancy.

## Special Considerations

- ◆ Teach the patient and family about retinitis pigmentosa.
- ◆ Encourage the patient to use sunglasses to protect the retina from ultraviolet light and to help preserve vision.
- ◆ Explain that the disorder is hereditary, and suggest genetic counseling for adults who risk transmitting it to their children.
- ◆ Encourage annual eye examinations to monitor the progress of the disease.

- ◆ Warn the patient of the potential for not being able to drive a car safely at night.
- ◆ Refer the patient to a social service agency or to the National Retinitis Pigmentosa Foundation for information and for counseling to prepare for eventual blindness.
- ◆ Because the prospect of blindness is frightening, it's important that you provide emotional support and guidance.



**PREVENTION** Encourage the patient to seek genetic counseling.

## Miscellaneous Disorders

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### OPTIC ATROPHY

Optic atrophy, or degeneration of the optic nerve, can develop spontaneously (primary) or can follow inflammation or edema of the nerve head (secondary). Some forms of this condition may subside without treatment, but degeneration of the optic nerve is irreversible.

### Pathophysiology

Optic atrophy usually results from central nervous system disorders (such as chiasmal tumors, syphilis, ischemic optic neuropathy, drugs, retinal vascular disease, or degenerative disease) or from end-stage glaucoma. Other causes include retinitis pigmentosa; chronic papilledema and papillitis; trauma; central retinal artery or vein occlusion that interrupts the blood supply to the optic nerve, causing degeneration of ganglion cells; ingestion of toxins, such as methanol and quinine; and deficiencies of vitamin B<sub>12</sub>, amino acids, and zinc.

Several rare forms of hereditary optic atrophy can affect children and young adults.

### Complications

- ◆ Central vision loss
- ◆ Peripheral vision loss
- ◆ Loss of contrast
- ◆ Loss of color vision

- ◆ Loss of visual field

## Signs and Symptoms

Optic atrophy causes abrupt or gradual painless loss of visual field or visual acuity, with changes in color vision.

## Diagnosis

**Dx CONFIRMING DIAGNOSIS** Visual acuity testing reveals poor vision. An afferent pupillary defect is noted when pupils are examined. Fundus examination through a dilated pupil with an ophthalmoscope shows pallor of the nerve head from loss of microvascular circulation in the disk and deposit of fibrous or glial tissue. Visual field testing reveals a scotoma and, possibly, major visual field impairment.

## Treatment

Optic atrophy is irreversible, so treatment aims to correct the underlying cause and prevent further vision loss. If a space-occupying lesion is the cause, neurosurgery may be required. In multiple sclerosis, optic neuritis often subsides spontaneously but may recur and improve repeatedly.

## Special Considerations

- ◆ Provide symptomatic care during diagnostic procedures and treatment.  
Assist the patient who's visually compromised to perform daily activities.
- ◆ Explain procedures, to minimize anxiety. Offer emotional support to help the patient deal with loss of vision.

## EXTRAOCULAR MOTOR NERVE PALSIIES

Extraocular motor nerve palsies are dysfunctions of the third, fourth, and sixth cranial nerves. The oculomotor (third cranial) nerve innervates the inferior, medial, and superior rectus muscles; the inferior oblique extraocular muscles; the pupilloconstrictor muscles; and the levator palpebrae muscles. The trochlear (fourth cranial) nerve innervates the superior oblique muscles. The abducens (sixth cranial) nerve innervates the lateral rectus muscles.

## **Pathophysiology**

Causes of these disorders vary, depending on the cranial nerve involved:

- ◆ Third nerve (oculomotor) palsy (acute ophthalmoplegia) may be congenital or acquired. Causes include intracranial tumors or aneurysms, diabetic neuropathy, and trauma.
- ◆ Fourth nerve (trochlear) palsy is most commonly caused by trauma.
- ◆ Sixth nerve (abducens) palsy commonly has an unknown etiology. Strokes are a common cause. Brainstem lesions, elevated intracranial pressure, inflamed petrous pyramid due to otitis media, cavernous sinus, orbital involvement with tumor and inflammation, or thyroid eye disease may be responsible for sixth nerve palsy.

## **Complications**

- ◆ Loss of eye muscle control (strabismus)
- ◆ Double vision (diplopia)
- ◆ Ptosis
- ◆ Dilated pupil (mydriasis)

## **Signs and Symptoms**

The most characteristic clinical symptom of extraocular motor nerve palsies is diplopia of recent onset, which varies in different visual fields, depending on the muscles affected.

Typically, the patient with third nerve palsy exhibits ptosis, exotropia (eye looks outward), pupil dilation, and unresponsiveness to light; the eye is unable to move and can't accommodate.

The patient with fourth nerve palsy displays diplopia and an inability to rotate the eye downward or upward. The head is usually turned to the side opposite the involved eye in superior oblique palsy to compensate for the diplopia.

Sixth nerve palsy causes one eye to turn; the eye can't abduct beyond the midline. To compensate for diplopia, the patient turns the head to the unaffected side and can develop torticollis.

## **Diagnosis**

Diagnosis necessitates an orthoptic examination to isolate the involved muscle, a complete neuro-ophthalmologic examination, and a thorough

patient history. Differential diagnosis of third, fourth, or sixth nerve palsy depends on the specific motor defect exhibited by the patient.

For all extraocular motor nerve palsies, a computed tomography scan or magnetic resonance imaging rules out tumors and may help detect the cause of the palsy, such as the cause of increased intracranial pressure. The patient is also evaluated for an aneurysm or diabetes. If sixth nerve palsy results from infection, culture and sensitivity tests identify the causative organism, and specific antibiotic therapy can be determined.

## Treatment

Identification of the underlying cause is essential because treatment for extraocular motor nerve palsies varies accordingly. Neurosurgery is necessary if the cause is a brain tumor or an aneurysm. For infection, massive I.V. doses of antibiotics may be appropriate.

## Special Considerations

- ♦ If the palsy results from thyroid eye disease, the patient must have normal thyroid levels before eye muscle surgery is attempted.

## GLAUCOMA

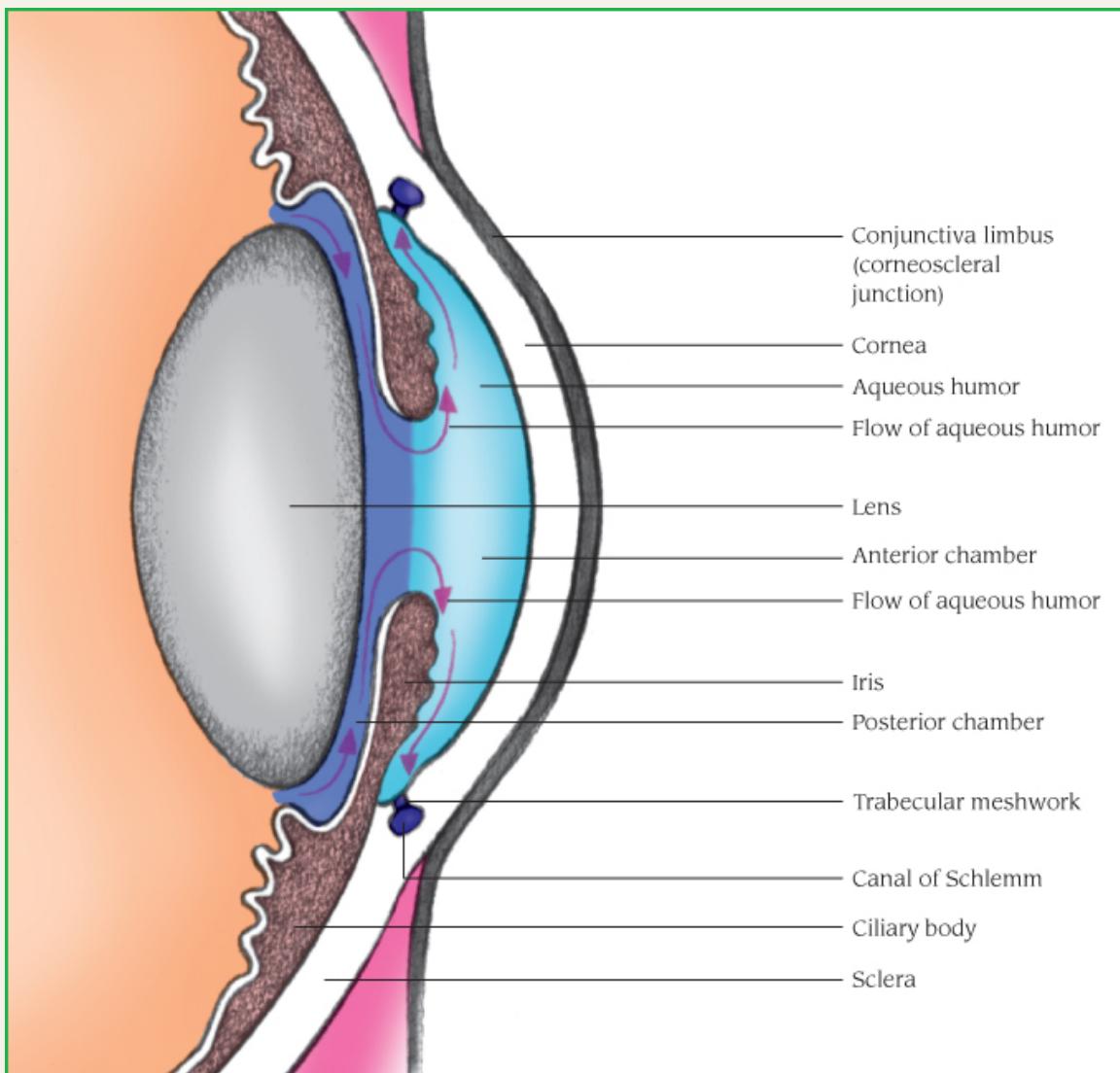
Glaucoma is a group of disorders characterized by damage to the optic nerve, most commonly associated with high IOP. If untreated, it can lead to gradual peripheral vision loss and, ultimately, blindness. (See *Blindness*, page 622.) Glaucoma occurs in several forms: chronic open-angle (primary), acute angle-closure, congenital (inherited as an autosomal recessive trait), and secondary to other causes. The prognosis for maintaining vision is good with early treatment.

## Pathophysiology

Chronic open-angle glaucoma results from overproduction of aqueous humor or obstruction to its outflow through the trabecular meshwork or the canal of Schlemm. (See *Normal flow of aqueous humor*.) This form of glaucoma, which is estimated to be present in 1% to 2% of people older than age 40, is frequently familial in origin and affects 90% of all patients with glaucoma. Diabetes and systemic hypertension have also been associated with this form of glaucoma.

## Normal Flow Of Aqueous Humor

Aqueous humor, a plasmalike fluid produced by the ciliary epithelium of the ciliary body, flows from the posterior chamber to the anterior chamber through the pupil. Here it flows peripherally and filters through the trabecular meshwork to the canal of Schlemm, through which the fluid ultimately enters venous circulation.



Acute angle-closure (narrow-angle) glaucoma results from obstruction to the outflow of aqueous humor due to anatomically narrow angles between the anterior iris and the posterior corneal surface, shallow anterior chambers, a thickened iris that causes angle closure on pupil dilation, or a bulging iris that presses on the trabeculae, closing the angle (peripheral anterior synechiae).

Persons of African descent are four times more likely to have this disorder than Caucasians, and people with a family history of open-angle glaucoma are twice as likely to develop it than people without a family history of this disorder. The use of systemic anticholinergic medications, such as atropine or eye dilation drops, in a person who's already at high risk for acute glaucoma increases the risk. Other risk factors include farsightedness and age-related changes that create an increase in IOP.

## Blindness

Blindness affects 28 million people worldwide. In the United States, blindness is legally defined as visual acuity of 20/200 or less in the better eye after best correction, or a visual field of 20 degrees or less in the better eye.

According to the World Health Organization, the most common causes of preventable blindness worldwide are trachoma, cataracts, onchocerciasis (microfilarial infection transmitted by a blackfly and other species of *Simulium*), and xerophthalmia (dryness of conjunctiva and cornea from vitamin A deficiency).

In the United States, the most common causes of acquired blindness are glaucoma, age-related macular degeneration, and diabetic retinopathy. However, the incidence of blindness from glaucoma is decreasing owing to early detection and treatment. Rarer causes of acquired blindness include herpes simplex keratitis, cataracts, and retinal detachment.

Congenital glaucoma occurs when there is an abnormal fluid drainage angle of the eye. It may be caused by congenital infections such as TORCH virus (toxoplasmosis, other [varicella, mumps, parvovirus, human

immunodeficiency virus], rubella, cytomegalovirus, and herpes), Sturge–Weber syndrome, or retinopathy of prematurity.

Secondary glaucoma can result from uveitis, trauma, or drugs (such as steroids). Neovascularization in the angle can result from vein occlusion or diabetes.

## Complications

- ◆ Blindness
- ◆ Vision loss

## Signs and Symptoms

Chronic open-angle glaucoma is usually bilateral, with insidious onset and a slowly progressive course. Symptoms appear late in the disease and include mild aching in the eyes, loss of peripheral vision, seeing halos around lights, and reduced visual acuity (especially at night) that isn't correctable with glasses.

Acute angle-closure glaucoma typically has a rapid onset, constituting an ophthalmic emergency. Symptoms include acute pain in a unilaterally inflamed eye, with pressure over the eye, moderate pupil dilation that's nonreactive to light, a cloudy cornea, blurring and decreased visual acuity, photophobia, and seeing halos around lights. Increased IOP may induce nausea and vomiting, which may cause glaucoma to be misinterpreted as GI distress. Unless treated promptly, this acute form of glaucoma produces blindness in 3 to 5 days.

## Diagnosis



**CONFIRMING DIAGNOSIS** *Loss of peripheral vision and disk changes confirm that glaucoma is present. Diagnosis is made by:*

- ◆ *testing IOP*
- ◆ *measuring the visual field and noting changes, such as loss of peripheral visual field*
- ◆ *observing changes in the cup/disk ratio of the optic nerve head (See Optic disk changes in glaucoma, page 623.)*

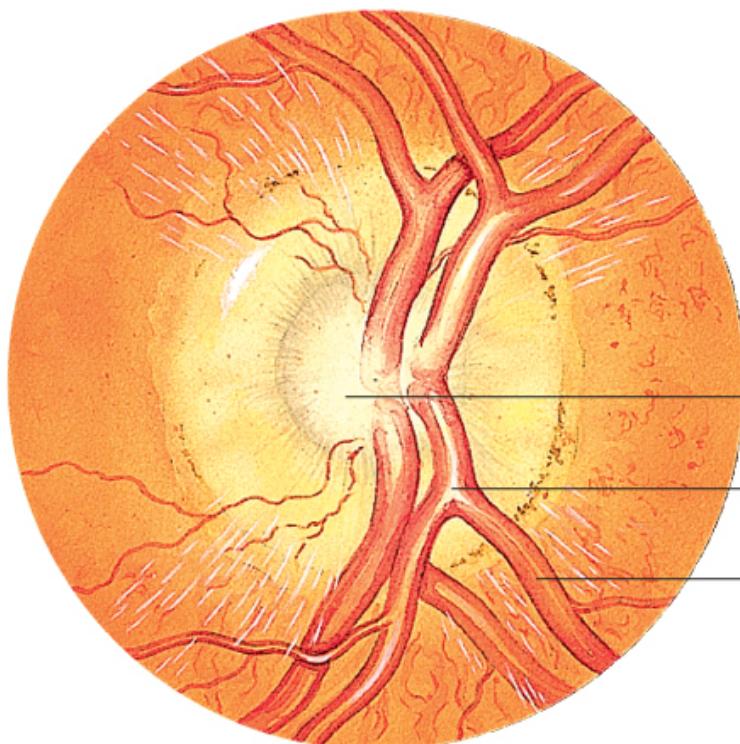


## **PATHOPHYSIOLOGY**

### **OPTIC DISK CHANGES IN GLAUCOMA**

Diagnosis of glaucoma is confirmed with a loss of peripheral vision and disk changes.

**NORMAL OPTIC DISK**

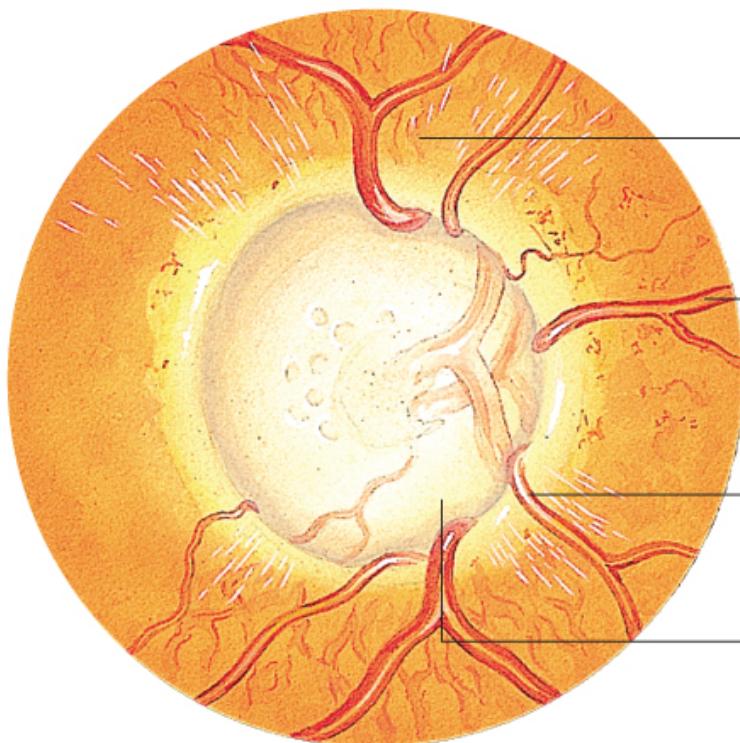


Optic disk

Central retinal artery and vein

Inferior macular artery and vein

**DISK CHANGES**



Decreased blood supply to retina

Blood vessels displaced nasally

Enlarged physiologic cup

Relevant diagnostic tests include:

- ◆ Tonometry (using an applanation tonometer [Tonopen] or air puff tonometer)—This test measures the IOP and provides a baseline for reference. Normal IOP ranges from 8 to 21 mm Hg. However, patients who fall within this normal range can develop signs and symptoms of glaucoma, and patients who have abnormally high pressure may have no clinical effects. Fingertip tension is another way to measure IOP. On gentle palpation of closed eyelids, one eye feels harder than the other in acute angle-closure glaucoma.
- ◆ Slit-lamp examination—The slit lamp facilitates examination of the anterior structures of the eye: the cornea, iris, and lens.
- ◆ Gonioscopy—By determining the angle of the anterior chamber of the eye, this test enables differentiation between chronic open-angle glaucoma and acute angle-closure glaucoma. The angle is normal in chronic open-angle glaucoma. However, in older patients, partial closure of the angle may occur, so that two forms of glaucoma may coexist.
- ◆ Ophthalmoscopy—This test enables the examiner to look at the fundus to establish if there are any cup/disk ratio changes. These changes appear later in chronic glaucoma if the disease isn't brought under control.
- ◆ Fundus photography—Pictures of the optic nerve head are made to track changes.
- ◆ Perimetry or visual field tests—These reveal the extent of damage to the optic neurons, signaled by an enlarged blind spot and loss of peripheral vision.

## Treatment

For chronic open-angle glaucoma, treatment initially decreases IOP through the use of one of five classes of drops; alpha antagonists such as brimonidine tartrate, beta blockers such as timolol (contraindicated for asthmatics or patients with bradycardia), prostaglandin analogs (such as latanoprost), carbonic anhydrase inhibitors (topical or oral), or miotics (such as pilocarpine).

Patients who are unresponsive to drug therapy may be candidates for argon laser trabeculoplasty (ALT) or a surgical filtering procedure called *trabeculectomy*, which creates an opening for aqueous outflow. In ALT, an

argon laser beam is focused on the trabecular meshwork of an open angle. This produces a thermal burn that changes the surface of the meshwork and increases the outflow of aqueous humor. In trabeculectomy, a flap of sclera is dissected free to expose the trabecular meshwork. Then this discrete tissue block is removed and a peripheral iridectomy is performed. This produces an opening for aqueous outflow under the conjunctiva, creating a filtering bleb. In chronic refractory glaucoma, a tube shunt or valve is used to keep IOP within normal limits.

Acute angle-closure glaucoma is an ocular emergency requiring immediate treatment to lower the high IOP. Drug therapy to lower IOP includes I.V. acetazolamide, pilocarpine (constricts the pupil, forcing the iris away from the trabeculae, allowing fluid to escape), timolol, and a topical steroid to quiet the inflammatory response, along with I.V. mannitol (20%) or oral glycerin (50%) to force fluid from the eye by making the blood hypertonic. Oral medication or topical drops may be prescribed separately or in combination. Severe pain may necessitate administration of opioid analgesics. If pressure doesn't decrease with drug therapy, laser iridotomies or surgical peripheral iridectomy must be performed promptly to save the patient's vision. Iridectomy relieves pressure by excising part of the iris to reestablish aqueous humor outflow. A prophylactic iridectomy is performed a few days later on the other eye to prevent an acute episode of glaucoma in the normal eye.

## Special Considerations

- ◆ Stress the importance of meticulous compliance with prescribed drug therapy to prevent an increase in IOP, resulting in disk changes and loss of vision.
- ◆ For the patient with acute angle-closure glaucoma, give medications as ordered, and prepare the patient physically and psychologically for laser iridotomies or surgery.
- ◆ Postoperative care after peripheral iridectomy includes cycloplegic eyedrops to relax the ciliary muscle and to decrease inflammation, thus preventing adhesions.
- ◆ Encourage ambulation immediately after surgery.
- ◆ Postoperative care after surgical filtering includes dilation and topical steroids to rest the pupil and topical steroids.

- ◆ Stress the importance of glaucoma screening for early detection and prevention. All people older than age 35, especially those with family histories of glaucoma, should have an annual tonometric examination.



**ALERT** Cycloplegics must be used only in the affected eye. The use of these drops in the normal eye may precipitate an attack of acute angle-closure glaucoma in this eye, threatening the patient's residual vision.

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# 13

## Ear, Nose, and Throat Disorders

### **Introduction**

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Ear, nose, and throat disorders rarely prove fatal (except for those resulting from neoplasms, epiglottitis, and neck trauma), but they may cause serious social, cosmetic, and communication problems. Untreated hearing loss or deafness can drastically impair the ability to interact with society. Ear disorders also can cause impaired equilibrium. Nasal disorders can cause changes in facial features and interfere with breathing and tasting. Diseases arising in the throat may threaten airway patency and interfere with speech. In addition, these disorders can cause considerable discomfort and pain for the patient and require thorough assessment and prompt treatment.

### **THE EAR**

Hearing begins when sound waves reach the tympanic membrane, which then vibrates the ossicles, incus, malleus, and stapes in the middle ear cavity. The stapes transmits these vibrations to the perilymphatic fluid in the inner ear by vibrating against the oval window. The vibrations then pass across the cochlea's fluid receptor cells in the basilar membrane, stimulating movement of the hair cells of the organ of Corti. The axons of the cochlear nerve terminate around the bases of those hair cells. Sound waves, which initiate impulses, travel over the auditory nerve (made up of

the cochlear nerve and the vestibular nerve) to the temporal lobe of the brain.

The inner ear structures also maintain the body's equilibrium and balance through the fluid in the semicircular canals. This fluid is set in motion by body movement and stimulates nerve cells that line the canals. These cells, in turn, transmit impulses to the cerebellum of the brain by way of the vestibular branch of the eighth cranial nerve (the acoustic nerve).

Although the ear can respond to sounds that vibrate at frequencies from 20 to 20,000 Hz, the range of normal speech is from 250 to 4,000 Hz, with 70% falling between 500 and 2,000 Hz. The ratio between sound intensities, the decibel (dB) is the unit for expressing the relative intensity (loudness) of sounds. A faint whisper registers 10 to 15 dB; average conversation, 50 to 60 dB; a shout, 85 to 90 dB. Hearing damage may follow exposure to sounds louder than 90 dB.

## ASSESSMENT

After obtaining a thorough patient history of any ear disease, inspect the auricle and surrounding tissue for deformities, lumps, and skin lesions. (See *Structures of the external ear*, page 626.) Ask the patient if they have ear pain. If you see inflammation, check for tenderness by moving the auricle and pressing on the tragus and the mastoid process. Check the ear canal for excessive cerumen, discharge, or foreign bodies.

### Structures of the External Ear

The structures of the external ear are depicted below.

### EXTERNAL EAR

Helix

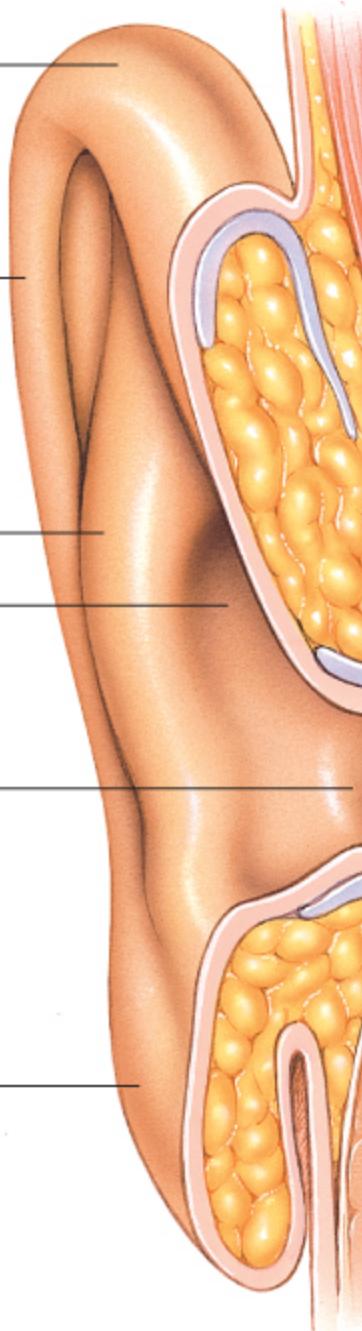
Darwinian  
tubercle

Anthelix

Concha

External  
acoustic  
meatus

Lobule of  
auricle



Ask the patient if they have had episodes of vertigo or blurred vision. To test for vertigo, have the patient stand on one foot and close their eyes, or

have them walk a straight line with their eyes closed. Ask them if they always fall to the same side and if the room seems to be spinning.

## AUDIOMETRIC TESTING

Audiometric testing evaluates hearing and determines the type and extent of hearing loss. The simplest but least reliable method for judging hearing acuity consists of covering one of the patient's ears, standing 18" to 24" (46 to 61 cm) from the uncovered ear, and whispering a short phrase or series of numbers. (Block the patient's vision to prevent lip reading.) Then ask the patient to repeat the phrase or series of numbers. To test hearing at both high and low frequencies, repeat the test in a normal speaking voice. (As an alternative, you can hold a ticking watch to the patient's ear.)

If you identify a hearing loss, further testing is necessary to determine if the loss is conductive or sensorineural. A conductive loss can result from faulty bone conduction (inability of the eighth cranial nerve to respond to sound waves traveling through the skull) or faulty air conduction (impaired transmission of sound through ear structures to the auditory nerve and, ultimately, the temporal lobe of the brain).

Sensorineural hearing loss results from damage to the cochlear or vestibulocochlear nerve, which can result from aging and prolonged exposure to high frequency or loud noises.

The following tests assess bone and air conduction:

- ◆ Impedance audiometry detects middle ear pathology, precisely determining the degree of tympanic membrane and middle ear mobility. One end of the impedance audiometer, a probe with three small tubes, is inserted into the external canal; the other end is attached to an oscillator. One tube delivers a low tone of variable intensity, the second contains a microphone, and the third, an air pump. A mobile tympanic membrane reflects minimal sound waves and produces a low-voltage curve on the graph. A tympanic membrane with decreased mobility reflects maximal sound waves and produces a high-voltage curve.
- ◆ Pure tone audiometry uses an audiometer to produce a series of pure tones of calibrated decibels of loudness at different frequencies (125 to 8,000 Hz). These test tones are conveyed to the patient's ears through headphones or a bone conduction (sound) vibrator. Speech threshold represents the loudness at which a person with normal hearing can

perceive the tone. Both air conduction and bone conduction are measured for each ear, and the results are plotted on a graph. If hearing is normal, the line is plotted at 0 dB. In adults, normal hearing may range from 0 to 25 dB.

- ◆ In the Rinne test, the base of a lightly vibrating tuning fork is placed on the mastoid process (bone conduction). Then the fork is moved to the front of the meatus, where the patient should continue to hear the vibrations (air conduction). The patient must determine which sounds are heard longer. In a positive Rinne test, sounds heard through air conduction are heard relatively longer than those heard through bone conduction. This may suggest sensorineural hearing loss. In a negative Rinne test, sounds heard through bone conduction are heard longer than those heard through air conduction, which may suggest a conductive loss.
- ◆ Speech audiometry uses the same technique as pure tone audiometry, but with speech, instead of pure tones, transmitted through the headset. (A person with normal hearing can hear and repeat 88% to 100% of transmitted words.)
- ◆ Tympanometry, using the impedance audiometer, measures tympanic membrane compliance with air pressure variations in the external canal and determines the degree of negative pressure in the middle ear.
- ◆ In Weber test (used for testing unilateral hearing loss), the handle of a lightly vibrating tuning fork is placed on the midline of the forehead. Normally, the patient should hear sounds equally in both ears. With conductive hearing loss, sound lateralizes (localizes) to the ear with the poorest hearing. With sensorineural loss, sound lateralizes to the better functioning ear.

## THE NOSE

As air travels between the septum and the turbinates, it touches sensory hairs (cilia) in the mucosal surface, which then add, retain, or remove moisture and particles in the air to ensure delivery of humid, bacteria-free air to the pharynx and lungs. In addition, when air touches the mucosal cilia, the resultant stimulation of the first cranial nerve sends nerve impulses to the olfactory area of the frontal cortex, providing the sense of smell.

## ASSESSMENT

Check the external nose for redness, edema, masses, or poor alignment. Marked septal cartilage depression may indicate saddle deformity because of septal destruction from trauma or congenital syphilis; extreme lateral deviation may result from injury. Red nostrils may indicate frequent nose blowing caused by allergies or infectious rhinitis. Dilated, engorged blood vessels may suggest alcoholism or constant exposure to the elements. A bulbous, discolored nose may be a sign of rosacea.

With a nasal speculum and adequate lighting, check nasal mucosa for pallor and edema or redness and inflammation, dried mucous plugs, furuncles, and polyps. Also, look for abnormal appearance of the capillaries, boggy turbinates, and a deviated or perforated septum. Check for nasal discharge (assess color, consistency, and odor) and blood. Profuse, thin, watery discharge may indicate allergy or cold; excessive, thin, purulent discharge may indicate cold or chronic sinus infection.

Check for sinus inflammation by applying pressure to the nostrils, orbital rims, and cheeks. Pain after pressure applied above the upper orbital rims indicates frontal sinus irritation; pain after pressure applied to the cheeks, maxillary sinus irritation.

## THE THROAT

Parts of the throat include the pharynx, epiglottis, and larynx. The pharynx is the passageway for food to the esophagus and air to the larynx. The epiglottis (the lid of the larynx) diverts material away from the glottis during swallowing. The larynx produces sounds by vibrating expired air through the vocal cords. Changes in vocal cord length and air pressure affect pitch and voice intensity. The larynx also stimulates the vital cough reflex when a foreign body touches its sensitive mucosa.

## ASSESSMENT

Using a bright light and a tongue blade, inspect the patient's mouth and throat. Look for inflammation or white patches, and any irregularities on the tongue or throat. Make sure the patient's airway isn't compromised and also assess vital signs. Watch for and immediately report signs of respiratory distress (dyspnea, tachycardia, tachypnea, inspiratory stridor, restlessness, and nasal flaring) and changes in voice or in skin color, such as circumoral or nail bed cyanosis. Assess symmetry of the tongue as well as function of

the soft palate. The main diagnostic test used in throat assessment is a culture to identify the infective organism.

## External Ear

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### OTITIS EXTERNA

Otitis externa, inflammation of the skin of the external ear canal and auricle, may be acute or chronic. Also known as *external otitis* and *swimmer's ear*, it's most common in the summer. With treatment, acute otitis externa usually subsides within 7 days—although it may become chronic—and tends to recur.

### Causes and Incidence

Otitis externa usually results from bacteria, such as *Pseudomonas*, *Proteus vulgaris*, *Staphylococcus aureus*, and streptococci and, sometimes, from fungi, such as *Aspergillus niger* and *Candida albicans* (fungal otitis externa is most common in tropical regions). Occasionally, chronic otitis externa results from dermatologic conditions, such as seborrhea or psoriasis. Allergic reactions stemming from nickel or chromium earrings, chemicals in hair spray, cosmetics, hearing aids, and medications (such as sulfonamide and neomycin, which is commonly used to treat otitis externa) can also cause otitis externa.

Predisposing factors include:

- ◆ Swimming in contaminated water. (Cerumen creates a culture medium for the waterborne organism.)
- ◆ Cleaning the ear canal with a cotton swab, bobby pin, finger, or other foreign object. (This irritates the ear canal and, possibly, introduces the infecting microorganism.)
- ◆ Exposure to dust or hair-care products (such as hair spray or other irritants), which causes the patient to scratch the ear, excoriating the auricle and canal.
- ◆ Regular use of earphones, earplugs, or earmuffs, which trap moisture in the ear canal, creating a culture medium for infection (especially if earplugs don't fit properly).
- ◆ Chronic drainage from a perforated tympanic membrane.

- ◆ Perfumes or self-administered eardrops.

## Pathophysiology

Otitis externa can take an acute or a chronic form. Acute disease commonly results from bacterial or fungal overgrowth in an ear canal subjected to excess moisture or to local trauma. Chronic disease often is part of a more generalized dermatologic or allergic problem. Symptoms of early acute and most chronic disease include pruritus and local discomfort. If left untreated, acute disease can be followed by canal edema, discharge, and pain, and eventually by extra-canal manifestations. Topical application of an acidifying solution is usually adequate in treating early disease. An antimicrobial-containing ototopical is the preferred treatment for later-stage acute disease, and oral antibiotic therapy is reserved for advanced disease or those who are immunocompromised. Preventive measures reduce recurrences and typically involve minimizing ear canal moisture, trauma, or exposure to materials that incite local irritation or contact dermatitis.

## Complications

- ◆ Complete closure of the ear canal
- ◆ Significant hearing loss
- ◆ Otitis media
- ◆ Cellulitis
- ◆ Abscesses
- ◆ Stenosis

## Signs and Symptoms

Acute otitis externa characteristically produces moderate to severe pain that's exacerbated by manipulating the auricle or tragus, clenching the teeth, opening the mouth, or chewing. Its other clinical effects may include fever, foul-smelling discharge, crusting in the external ear, regional cellulitis, partial hearing loss, and itching. It's usually difficult to view the tympanic membrane because of pain in the external canal. Hearing acuity is normal unless complete occlusion has occurred.

Fungal otitis externa may be asymptomatic, although *A. niger* produces a black or gray, blotting, paper-like growth in the ear canal. In chronic otitis externa, pruritus replaces pain, and scratching may lead to scaling and skin thickening. Aural discharge may also occur.

## Diagnosis



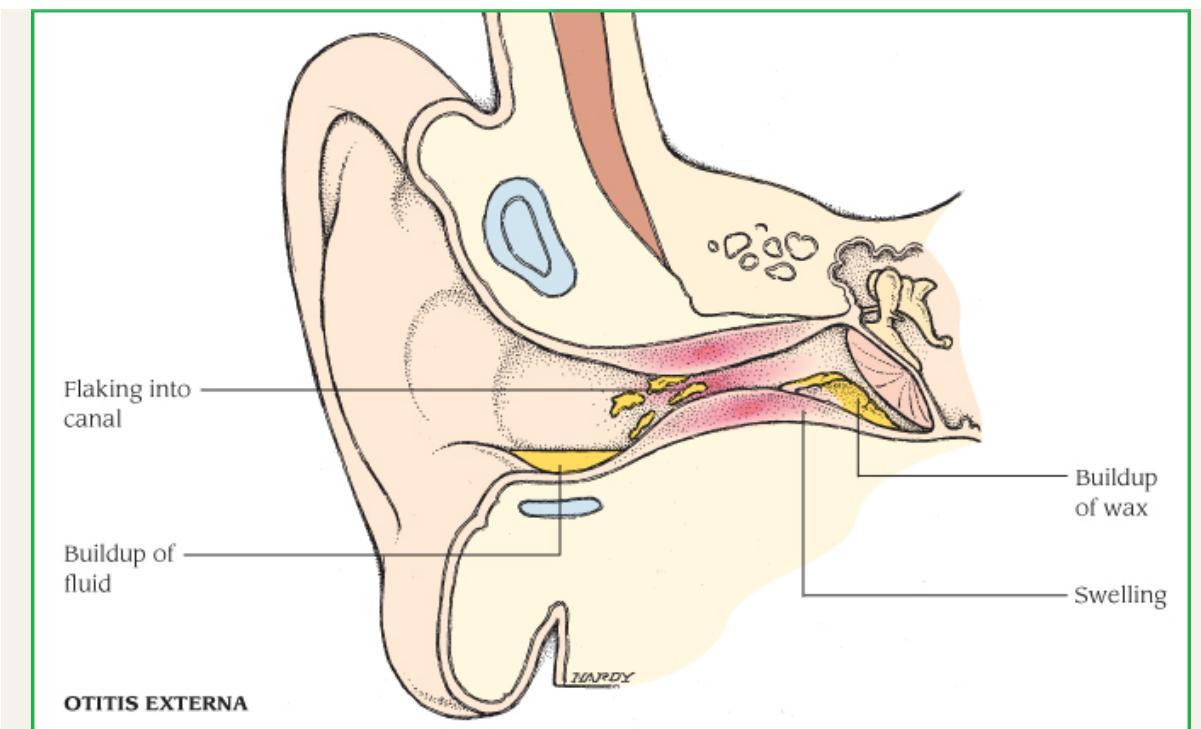
### CONFIRMING DIAGNOSIS

*Physical examination confirms otitis externa. In acute otitis externa, otoscopy reveals a swollen external ear canal (sometimes to the point of complete closure), preauricular lymphadenopathy (tender nodes anterior to the tragus, posterior to the ear, or in the upper neck), and, occasionally, regional cellulitis.*

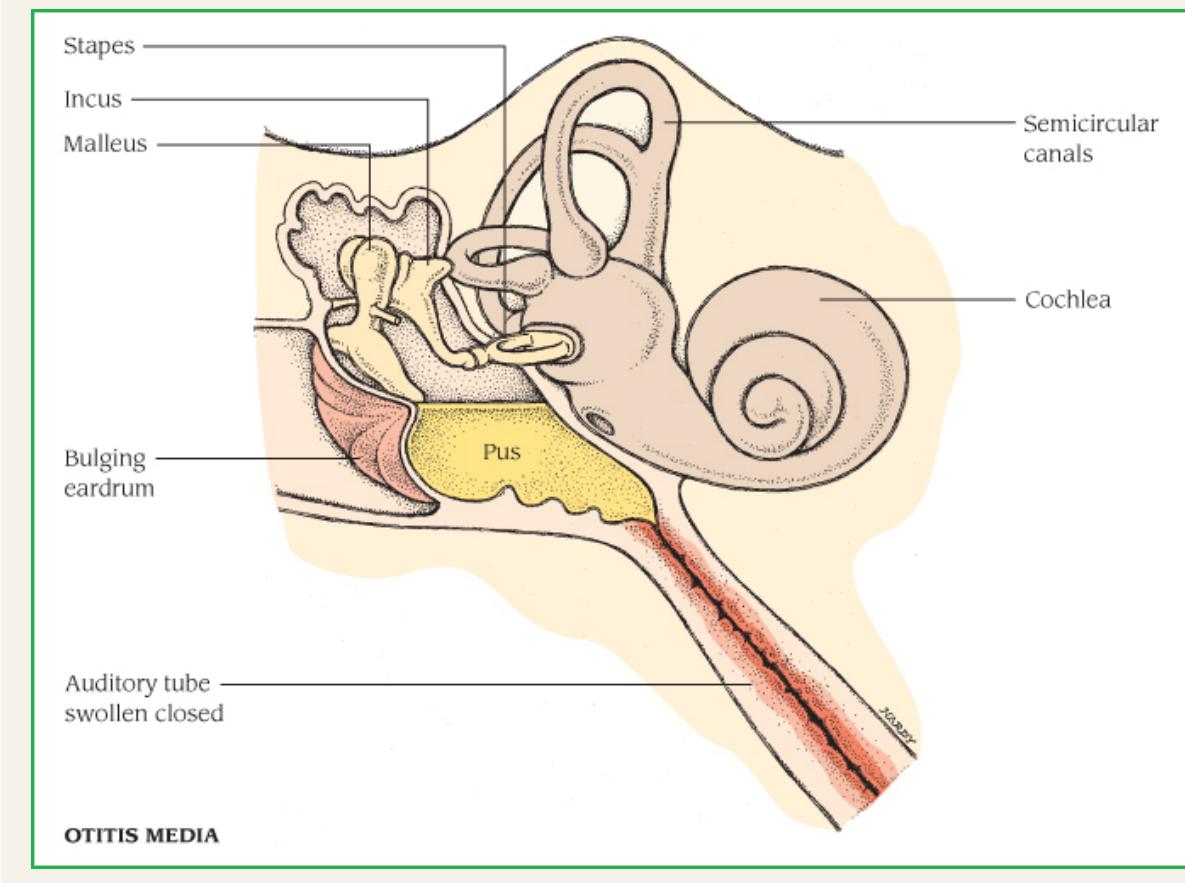
In fungal otitis externa, removal of the growth reveals thick red epithelium. Microscopic examination or culture and sensitivity tests can identify the causative organism and determine antibiotic treatment. Pain on palpation of the tragus or auricle distinguishes acute otitis externa from acute otitis media. (See *Differentiating acute otitis externa from acute otitis media*, page 629.)

## Differentiating Acute Otitis Externa From Acute Otitis Media

Use the assessment findings shown below to help differentiate acute otitis externa from acute otitis media.



Acute Otitis Externa (Occurs Primarily In Summer)



## Acute Otitis Media (Occurs Primarily In Winter)

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In chronic otitis externa, physical examination reveals thick red epithelium in the ear canal. Severe chronic otitis externa may reflect underlying diabetes mellitus, hypothyroidism, or nephritis. Microscopic examination or culture and sensitivity tests can identify the causative organism and help in the determination of antibiotic treatment.

### Treatment

To relieve the pain of acute otitis externa, treatment includes heat therapy to the preauricular region (heat lamp; hot, damp compresses; or a heating pad), aspirin or acetaminophen, and codeine. Instillation of antibiotic eardrops (with or without hydrocortisone) follows cleaning of the ear and removal of debris. However, a corticosteroid helps reduce the inflammatory response. If fever persists or regional cellulitis or tender postauricular adenopathy develops, a systemic antibiotic is necessary.

If the ear canal is too edematous for the instillation of eardrops, an ear wick may be used for the first few days.

Topical treatment is generally required for otitis externa, as systemic antibiotics alone aren't sufficient. Analgesics, such as acetaminophen or ibuprofen, may be required temporarily.

As with other forms of this disorder, fungal otitis externa necessitates careful cleaning of the ear. Application of a keratolytic or 2% salicylic acid in cream-containing nystatin may help treat otitis externa resulting from candidal organisms. Instillation of slightly acidic eardrops creates an unfavorable environment in the ear canal for most fungi as well as *Pseudomonas*. No specific treatment exists for otitis externa caused by *A. niger*, except repeated cleaning of the ear canal with baby oil.

In chronic otitis externa, primary treatment consists of cleaning the ear and removing debris. Supplemental therapy includes instillation of antibiotic eardrops or application of antibiotic ointment or cream (neomycin, bacitracin, or polymyxin B, possibly combined with hydrocortisone). Another ointment contains phenol, salicylic acid, precipitated sulfur, and petroleum jelly and produces exfoliative and antipruritic effects.

For mild chronic otitis externa, treatment may include instillation of antibiotic eardrops once or twice weekly and wearing of specially fitted earplugs while the patient is showering, shampooing, or swimming.

## Special Considerations

If the patient has acute otitis externa:

- ◆ The patient shouldn't participate in any swimming activity.
- ◆ Have the patient return to the clinic in 1 week for evaluation of the tympanic membrane to make sure it's intact.
- ◆ Monitor vital signs, particularly temperature. Watch for and record the type and amount of aural drainage.
- ◆ Remove debris and gently clean the ear canal with mild Burow solution (aluminum acetate). Place a wisp of cotton soaked with solution into the ear, and apply a saturated compress directly to the auricle. Afterward, dry the ear gently but thoroughly. (In severe otitis externa, such cleaning may be delayed until after initial treatment with antibiotic eardrops.)
- ◆ To instill eardrops in an adult, grasp the helix and pull upward and backward to straighten the canal.
- ◆ Tell the patient to notify the physician if they develop an allergic reaction to the antibiotic drops or ointment, which may be indicated by increased swelling and discomfort of the area and worsening of other symptoms.



**PEDIATRIC TIP** *To instill eardrops in a child, pull the earlobe downward and backward. To ensure that the drops reach the epithelium, insert a wisp of cotton moistened with eardrops.*

If the patient has chronic otitis externa, clean the ear thoroughly. Use wet soaks intermittently on oozing or infected skin. If the patient has a chronic fungal infection, clean the ear canal well, and then apply an exfoliative ointment.

- ◆ Urge prompt treatment for otitis media to prevent perforation of the tympanic membrane. (See *Preventing otitis externa*.)

## **PREVENTION**



### **PREVENTING OTITIS EXTERNA**

Any patient who has experienced otitis externa should be taught to prevent a recurrence by avoiding irritants, such as hair-care products and earrings, and by avoiding cleaning the ears with cotton-tipped applicators or other objects. Encourage the patient to keep water out of the ears when showering or shampooing by using lamb's wool earplugs, coated with petroleum jelly. Also, parents of young children should be told that modeling clay makes a tight seal to prevent water from getting into the external ear canal.

In addition, when the patient goes swimming, keep their head above water or wear earplugs. After swimming, the patient should instill one or two drops of a mixture that is one-half 70% alcohol and one-half white vinegar to toughen the skin of the external ear canal.



**ELDER TIP** *If the patient is an elderly person or has diabetes, evaluate for malignant otitis externa.*



**PEDIATRIC TIP** *Children who have an intact tympanic membrane but are predisposed to otitis externa from swimming should instill two to three drops of a 1:1 solution of white vinegar and 70% ethyl alcohol into their ears before and after swimming.*

## **BENIGN TUMORS OF THE EAR CANAL**

Benign tumors may develop anywhere in the ear canal. Common types include keloids, osteomas, and sebaceous cysts; their causes vary. (See *Causes and characteristics of benign ear tumors*.) These tumors seldom become malignant; with proper treatment, the prognosis is excellent.

### **Causes and Characteristics of Benign Ear Tumors**

| <b>Tumor</b> | <b>Causes and incidence</b> | <b>Characteristics</b> |
|--------------|-----------------------------|------------------------|
|--------------|-----------------------------|------------------------|

| <b>Tumor</b>   | <b>Causes and incidence</b>  | <b>Characteristics</b>   |
|----------------|--|--|
| Keloid         | <ul style="list-style-type: none"> <li>◆ Surgery or trauma such as ear piercing</li> <li>◆ Most common in blacks</li> </ul>  | <ul style="list-style-type: none"> <li>◆ Hypertrophy and fibrosis of scar tissue</li> <li>◆ Commonly recurs</li> </ul>   |
| Osteoma        | <ul style="list-style-type: none"> <li>◆ Idiopathic growth</li> <li>◆ Predisposing factor: swimming in cold water</li> <li>◆ Three times more common in males than in females</li> <li>◆ Seldom occurs before adolescence</li> </ul> | <ul style="list-style-type: none"> <li>◆ Bony outgrowth from wall of external auditory meatus</li> <li>◆ Usually bilateral and multiple (exostoses)</li> <li>◆ May be circumscribed or diffuse, nondisplaceable, nontender</li> </ul>  |
| Sebaceous cyst | <ul style="list-style-type: none"> <li>◆ Obstruction of a sebaceous gland</li> </ul>   | <ul style="list-style-type: none"> <li>◆ Painless, circumscribed, round mass of variable size filled with oily, fatty, glandular secretions</li> <li>◆ May occur on external ear and outer third of external auditory canal</li> </ul> |

## Signs and Symptoms

A benign ear tumor is usually asymptomatic, unless it becomes infected, in which case pain, fever, or inflammation may result. (Pain is usually a sign of a malignant tumor.) If the tumor grows large enough to obstruct the ear canal by itself or through accumulated cerumen and debris, it may cause hearing loss and the sensation of pressure.

## Diagnosis

 **CONFIRMING DIAGNOSIS** Clinical features and patient history suggest a benign tumor of the ear canal; otoscopy confirms it. To rule out cancer, a biopsy may be necessary.

## Treatment

Generally, a benign tumor requires surgical excision if it obstructs the ear canal, is cosmetically undesirable, or becomes malignant.

Treatment for keloids may include surgery followed by repeated injections of long-acting steroids into the suture line. Excision must be complete, but even this may not prevent recurrence.

Surgical excision of an osteoma consists of elevating the skin from the surface of the bony growth and shaving the osteoma with a mechanical burr or drill.

Before surgery, a sebaceous cyst requires preliminary treatment with antibiotics, to reduce inflammation. To prevent recurrence, excision must be complete, including the sac or capsule of the cyst.

## Special Considerations

Because treatment for benign ear tumors generally doesn't require hospitalization, focus care on emotional support and on providing appropriate patient education so that the patient follows the therapeutic plan properly when he's at home.

- ◆ Thoroughly explain diagnostic procedures and treatment to the patient and family. Reassure them and answer any questions they may have.
- ◆ After surgery, instruct the patient in good aural hygiene. Until the ear is completely healed, advise the patient not to insert anything into their ear or allow water to get into it. Suggest that they cover their ears with a cap when showering.
- ◆ Teach the patient how to recognize signs of infection, such as pain, fever, localized redness, and swelling. If the patient detects any of these signs, instruct to report them immediately.

## Middle Ear

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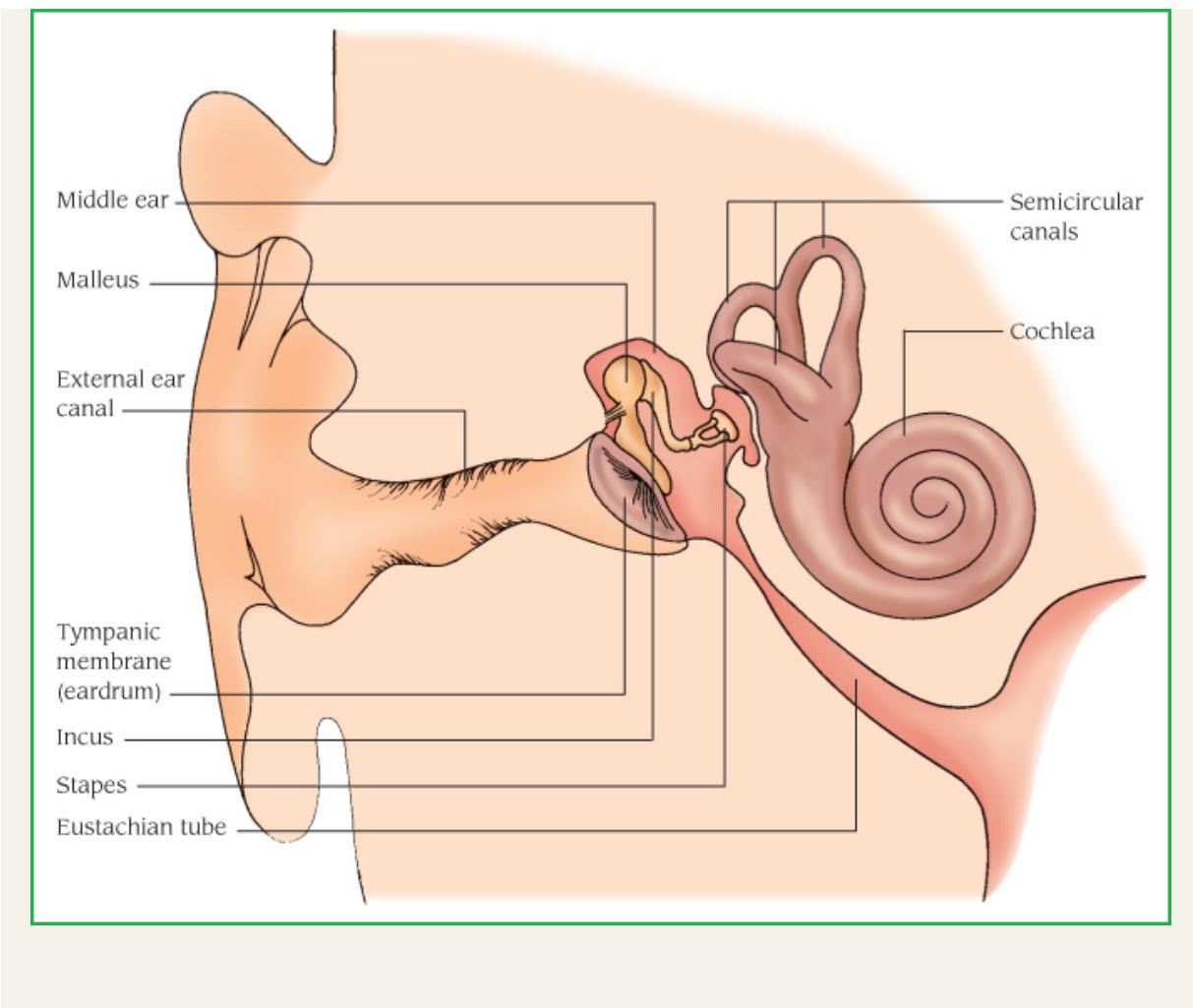
### OTITIS MEDIA

Otitis media, inflammation of the middle ear, may be suppurative or secretory, acute, persistent, unresponsive, or chronic. With prompt treatment, the prognosis for acute otitis media is excellent; however, prolonged accumulation of fluid within the middle ear cavity causes chronic otitis media and, possibly, perforation of the tympanic membrane. (See *Site of otitis media*.)

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#### Site of Otitis Media

The common site of otitis media is shown below.



Chronic suppurative otitis media may lead to scarring, adhesions, and severe structural or functional ear damage. Chronic secretory otitis media, with its persistent inflammation and pressure, may cause conductive hearing loss.

Recurrent otitis media is defined as three near-acute otitis media episodes within 6 months or four episodes of acute otitis media within 1 year.

Otitis media with complications involves damage to middle ear structures (such as adhesions, retraction, pockets, cholesteatoma, and intratemporal and intracranial complications).

## Causes and Incidence

Otitis media results from disruption of eustachian tube patency. In the suppurative form, respiratory tract infection, allergic reaction, nasotracheal intubation, or positional changes allow nasopharyngeal flora to reflux

through the eustachian tube and colonize the middle ear. Suppurative otitis media usually results from bacterial infection with pneumococcus, *Haemophilus influenzae* (the most common cause in children younger than age 6), *Moraxella catarrhalis*, beta-hemolytic streptococci, staphylococci (most common cause in children age 6 or older), or gram-negative bacteria. Predisposing factors include the normally wider, shorter, more horizontal eustachian tubes and increased lymphoid tissue in children, as well as anatomic anomalies. Chronic suppurative otitis media results from inadequate treatment for acute otitis episodes or from infection by resistant strains of bacteria or, rarely, tuberculosis.

Secretory otitis media results from obstruction of the eustachian tube. This causes a buildup of negative pressure in the middle ear that promotes transudation of sterile serous fluid from blood vessels in the membrane of the middle ear. Such effusion may be secondary to eustachian tube dysfunction from viral infection or allergy. It may also follow barotrauma (pressure injury caused by the inability to equalize pressures between the environment and the middle ear), as occurs during rapid aircraft descent in a person with an upper respiratory tract infection (URTI) or during rapid underwater ascent in scuba diving (barotitis media).

Chronic secretory otitis media follows persistent eustachian tube dysfunction from mechanical obstruction (adenoidal tissue overgrowth or tumors), edema (allergic rhinitis or chronic sinus infection), or inadequate treatment for acute suppurative otitis media.

Acute otitis media is common in children; its incidence rises during the winter months, paralleling the seasonal rise in nonbacterial respiratory tract infections. Chronic secretory otitis media most commonly occurs in children with tympanostomy tubes or those with a perforated tympanic membrane.

## **Pathophysiology**

Acute otitis media is an acute infection of the middle ear, usually lasting less than 6 weeks. The primary cause of acute otitis media is usually *Streptococcus pneumoniae*, *H. influenzae*, and *M. catarrhalis*, which enter the middle ear after eustachian tube dysfunction caused by obstruction related to upper respiratory infections, inflammation of surrounding structures (e.g., sinusitis, adenoid hypertrophy), or allergic reactions (e.g., allergic rhinitis). Bacteria can enter the eustachian tube from contaminated

secretions in the nasopharynx and the middle ear from a tympanic membrane perforation. A purulent exudate is usually present in the middle ear, resulting in a conductive hearing loss.

## Complications

- ◆ Spontaneous rupture of the tympanic membrane
- ◆ Persistent perforation
- ◆ Chronic otitis media
- ◆ Mastoiditis
- ◆ Abscesses
- ◆ Vertigo
- ◆ Permanent hearing loss

## Signs and Symptoms

Clinical features of acute suppurative otitis media include severe, deep, throbbing pain (from pressure behind the tympanic membrane); signs of URTI (sneezing or coughing); mild to very high fever; hearing loss (usually mild and conductive); tinnitus; dizziness; nausea; and vomiting. Other possible effects include bulging of the tympanic membrane, with concomitant erythema, and purulent drainage in the ear canal from tympanic membrane rupture. However, many patients are asymptomatic.

Acute secretory otitis media produces a severe conductive hearing loss—which varies from 15 to 35 dB, depending on the thickness and amount of fluid in the middle ear cavity—and, possibly, a sensation of fullness in the ear and popping, crackling, or clicking sounds on swallowing or with jaw movement. Accumulation of fluid may also cause the patient to hear an echo when the patient speaks and to experience a vague feeling of top-heaviness.

The cumulative effects of chronic otitis media include thickening and scarring of the tympanic membrane, decreased or absent tympanic membrane mobility, cholesteatoma (a cystlike mass in the middle ear), and, in chronic suppurative otitis media, a painless, purulent discharge. The extent of associated conductive hearing loss varies with the size and type of tympanic membrane perforation and ossicular destruction.

If the tympanic membrane has ruptured, the patient may state that the pain has suddenly stopped. Complications may include abscesses (brain, subperiosteal, and epidural), sigmoid sinus or jugular vein thrombosis,

septicemia, meningitis, suppurative labyrinthitis, facial paralysis, and otitis externa.



**PEDIATRIC TIP** *The following factors increase a child's risk of developing otitis media:*

- ◆ *acute otitis media in the first year after birth (recurrent otitis media)*
- ◆ *day care*
- ◆ *family history of middle ear disease*
- ◆ *formula feeding*
- ◆ *male gender*
- ◆ *sibling history of otitis media*
- ◆ *smoking in the household*

Acute otitis media may not produce any symptoms in the first few months of life; irritability may be the only indication of earache.

## Diagnosis

In acute suppurative otitis media, otoscopy reveals obscured or distorted bony landmarks of the tympanic membrane. Pneumatoscopy can show decreased tympanic membrane mobility, but this procedure is painful with an obviously bulging, erythematous tympanic membrane. The pain pattern is diagnostically significant: For example, in acute suppurative otitis media, pulling the auricle *doesn't* exacerbate the pain. A culture of the ear drainage identifies the causative organism.

In acute secretory otitis media, otoscopic examination reveals tympanic membrane retraction, which causes the bony landmarks to appear more prominent.

Examination also detects clear or amber fluid behind the tympanic membrane. If hemorrhage into the middle ear has occurred, as in barotrauma, the tympanic membrane appears blue-black.

In chronic otitis media, patient history discloses recurrent or unresolved otitis media. Otoscopy shows thickening, sometimes scarring, and decreased mobility of the tympanic membrane; pneumatoscopy shows decreased or absent tympanic membrane movement. A history of recent air travel or scuba diving suggests barotitis media.

Tympanocentesis for microbiologic diagnosis is recommended for treatment failures and may be followed by myringotomy. Tympanometry, acoustic reflex measurement, or acoustic reflexometry may be needed to document the presence of fluid in the middle ear. White blood cell count is higher in bacterial otitis media than in sterile otitis media. Mastoid X-rays or computed tomography (CT) scan of the head or mastoids may show the spreading of the infection beyond the middle ear.

## Treatment

In acute suppurative otitis media, antibiotic therapy includes amoxicillin. In areas with a high incidence of beta-lactamase-producing *H. influenzae* and in patients who aren't responding to ampicillin or amoxicillin, amoxicillin/clavulanate potassium may be used. For those who are allergic to penicillin derivatives, therapy may include cefaclor or trimethoprim and sulfamethoxazole. Severe, painful bulging of the tympanic membrane usually necessitates myringotomy. Broad-spectrum antibiotics can help prevent acute suppurative otitis media in high-risk patients. A single dose of ceftriaxone 50 mg/kg is effective against major pathogens but is expensive and is reserved for very sick infants. In the patient with recurring otitis media, antibiotics must be used with discretion to prevent the development of resistant strains of bacteria.

In acute secretory otitis media, inflation of the eustachian tube using Valsalva maneuver several times a day may be the only treatment required. Otherwise, nasopharyngeal decongestant therapy may be helpful. It should continue for at least 2 weeks and, sometimes, indefinitely, with periodic evaluation. If decongestant therapy fails, myringotomy and aspiration of middle ear fluid are necessary, followed by insertion of a polyethylene tube into the tympanic membrane, for immediate and prolonged equalization of pressure. The tube falls out spontaneously after 9 to 12 months. Concomitant treatment for the underlying cause (such as elimination of allergens, or adenoidectomy for hypertrophied adenoids) may also be helpful in correcting this disorder.

Treatment for chronic otitis media includes broad-spectrum antibiotics, such as amoxicillin/clavulanate potassium or cefuroxime, for exacerbations of acute otitis media; elimination of eustachian tube obstruction; treatment for otitis externa; myringoplasty and tympanoplasty to reconstruct middle

ear structures when thickening and scarring are present; and, possibly, mastoidectomy. Cholesteatoma requires excision.

## Special Considerations

- ◆ Explain all diagnostic tests and procedures. After myringotomy, maintain drainage flow. Don't place cotton or plugs deeply into the ear canal; however, sterile cotton may be placed loosely in the external ear to absorb drainage. To prevent infection, change the cotton whenever it gets damp, and wash hands before and after giving ear care. Watch for and report headache, fever, severe pain, or disorientation.
- ◆ After tympanoplasty, reinforce dressings and observe for excessive bleeding from the ear canal. Administer analgesics as needed. Warn the patient against blowing the nose or getting the ear wet when bathing.
- ◆ Encourage the patient to complete the prescribed course of antibiotic treatment. If nasopharyngeal decongestants are ordered, teach correct instillation.
- ◆ Suggest application of heat to the ear to relieve pain. (See *Preventing otitis media*, page 635.)



### PREVENTION PREVENTING OTITIS MEDIA

For a patient recovering from otitis media at home, teach these guidelines to help prevent a recurrence.

Instruct the patient how to recognize upper respiratory infections and encourage early treatment. Encourage the patient to get a pneumococcal vaccine to prevent infections that can cause respiratory and aural infections.

Tell parents to wash children's toys and promote frequent hand washing. For infants, tell parents to avoid the use of pacifiers and encourage breast-feeding for at least the first 6 months of the child's life. It has been shown that breast milk contains antibodies that protect the infant from ear infections. If the child is bottle-fed, instruct the parents not to feed the infant in a supine position and not to put the child to bed with a bottle. Explain that doing so could

cause reflux of nasopharyngeal flora. Also, teach the parent to keep the child away from secondhand smoke.

To promote eustachian tube patency, instruct the patient to perform Valsalva maneuver several times a day, especially during airplane travel. Also, explain adverse reactions to the prescribed medications, emphasizing those that require immediate medical attention.

- ◆ Advise the patient with acute secretory otitis media to watch for and immediately report pain and fever—signs of secondary infection.
- ◆ Identify and treat allergies.

## MASTOIDITIS

Mastoiditis is a bacterial infection and inflammation of the air cells of the mastoid antrum. Although the prognosis is good with early treatment, possible complications include meningitis, facial paralysis, brain abscess, and suppurative labyrinthitis.

### Causes and Incidence

Bacteria that cause mastoiditis include pneumococci, *H. influenzae*, *M. catarrhalis*, beta-hemolytic streptococci, staphylococci, and gram-negative organisms. Mastoiditis is usually a complication of chronic otitis media; less frequently, it develops after acute otitis media. An accumulation of pus under pressure in the middle ear cavity results in necrosis of adjacent tissue and extension of the infection into the mastoid cells. Chronic systemic diseases or immunosuppression may also lead to mastoiditis. Anaerobic organisms play a role in chronic mastoiditis.



## PREVENTION PREVENTING OTITIS EXTERNA

Any patient who has experienced otitis externa should be taught to prevent a recurrence by avoiding irritants, such as hair-care products and earrings, and by avoiding cleaning the ears with cotton-tipped applicators or other objects. Encourage the patient to keep water out of the ears when showering or shampooing by using lamb's wool earplugs,

coated with petroleum jelly. Also, parents of young children should be told that modeling clay makes a tight seal to prevent water from getting into the external ear canal.

In addition, when the patient goes swimming, keep their head above water or wear earplugs. After swimming, the patient should instill one or two drops of a mixture that is one-half 70% alcohol and one-half white vinegar to toughen the skin of the external ear canal.



**PEDIATRIC TIP** *Acute otitis media increases a child's risk of developing mastoiditis. If mastoiditis does occur in infants younger than age 1, the swelling occurs superior to the ear and pushes the auricle downward instead of outward. I.V. antibiotic treatment choice includes ampicillin or cefuroxime. Before antibiotics, mastoiditis was one of the leading causes of death in children; now, it's uncommon and less dangerous.*

## Pathophysiology

Mastoiditis, inflammation of the mastoid process, a projection of the temporal bone just behind the ear. Mastoiditis, which primarily affects children, usually results from an infection of the middle ear (otitis media). Symptoms include pain and swelling behind the ear and over the side of the head and fever. An abscess may develop; this indicates that the infection has eroded the bone and destroyed its outer layer. Mastoiditis may affect other structures within the cranium and produce complications including meningitis, abscesses of the dura mater covering the brain; infection or blood clots of the lateral sinus (the large blood channel emptying into the internal jugular vein); and infection of the labyrinth (the inner ear) containing the balance and hearing apparatus. Mastoiditis is a rare condition that is treated by the early administration of antibiotics. Surgical drainage and removal of diseased bone may be necessary if antibiotics are not successful.

## Complications

- ◆ Destruction of the mastoid bone
- ◆ Facial paralysis
- ◆ Meningitis

- ◆ Partial or complete hearing loss

## Signs and Symptoms

Primary clinical features include a dull ache and tenderness in the area of the mastoid process, low-grade fever, headache, and a thick, purulent discharge that gradually becomes more profuse, possibly leading to otitis externa. Postauricular erythema and edema may push the auricle out from the head; pressure within the edematous mastoid antrum may produce swelling and obstruction of the external ear canal, causing conductive hearing loss.

## Diagnosis

X-rays or CT scan of the mastoid area reveal hazy mastoid air cells; the bony walls between the cells appear decalcified. Audiometric testing may reveal a conductive hearing loss. Physical examination shows a dull, thickened, and edematous tympanic membrane, if the membrane isn't concealed by obstruction. During examination, the external ear canal is cleaned; persistent oozing into the canal indicates perforation of the tympanic membrane.

## Treatment

Treatment for mastoiditis consists of intense parenteral antibiotic therapy. Reasonable initial antibiotic choices include ceftriaxone with nafcillin or clindamycin. If bone damage is minimal, myringotomy or tympanocentesis drains purulent fluid and provides a specimen of discharge for culture and sensitivity testing. Recurrent or persistent infection or signs of intracranial complications necessitate simple mastoidectomy. This procedure involves removal of the diseased bone and cleaning of the affected area, after which a drain is inserted.

A chronically inflamed mastoid requires radical mastoidectomy (excision of the posterior wall of the ear canal, remnants of the tympanic membrane, and the malleus and incus, although these bones are usually destroyed by infection before surgery). The stapes and facial nerve remain intact. Radical mastoidectomy, which is seldom necessary because of antibiotic therapy, doesn't drastically affect the patient's hearing because significant hearing loss precedes surgery. With either surgical procedure, the patient continues

oral antibiotic therapy for several weeks after surgery and facility discharge. The prognosis is good if treatment is started early.

Indications for immediate surgical intervention include meningitis, brain abscess, cavernous sinus thrombosis, acute suppurative labyrinthitis, and facial palsy.

## Special Considerations

- ◆ After simple mastoidectomy, give pain medication as needed. Check wound drainage and reinforce dressings (the surgeon usually changes the dressing daily and removes the drain in 72 hours). Check the patient's hearing, and watch for signs of complications, especially infection (either localized or extending to the brain); facial nerve paralysis, with unilateral facial drooping; bleeding; and vertigo, especially when the patient stands.
- ◆ After radical mastoidectomy, the wound is packed with petroleum gauze or gauze treated with an antibiotic ointment. Give pain medication before the packing is removed, on the fourth or fifth postoperative day.
- ◆ Because of stimulation to the inner ear during surgery, the patient may feel dizzy and nauseated for several days afterward. Keep the side rails up, and assist the patient with ambulation. Also, give antiemetics as needed.
- ◆ Before discharge, teach the patient and family how to change and care for the dressing. Urge compliance with the prescribed antibiotic treatment and promote regular follow-up care.
- ◆ If the patient is an elderly person or diabetic, evaluate for malignant otitis externa.



**ELDER TIP** Encourage the patient to seek early treatment for ear infections.

## OTOSCLEROSIS

The most common cause of chronic, progressive conductive hearing loss, otosclerosis is the slow formation of spongy bone in the otic capsule, particularly at the oval window. With surgery, the prognosis is good.

## Causes and Incidence

Otosclerosis appears to result from a genetic factor transmitted as an autosomal dominant trait; many patients report family histories of hearing loss (excluding presbycusis). Pregnancy may trigger onset of this condition.

Otosclerosis occurs in at least 10% of the U.S. population. It's three times more prevalent in females than in males, usually affecting people between ages 15 and 30. Whites are most susceptible.

## **Pathophysiology**

Otosclerosis is a localized disease of bone remodeling within the otic capsule of the human temporal bone. Unlike other similar bone diseases, it does not occur outside of the temporal bone. These lesions seem to begin by resorption of stable otic capsule bone in adults, followed by a reparative phase with bone deposition. There are clearly genetic factors that lead to this disease, but measles virus infection and autoimmunity also may play contributing roles. Surgical correction of the conductive hearing loss is highly effective, but nonsurgical intervention has not yet been shown to prevent or slow the disease.

## **Complications**

- ◆ Bilateral conductive hearing loss
- ◆ Taste disturbance

## **Signs and Symptoms**

Spongy bone in the otic capsule immobilizes the footplate of the normally mobile stapes, disrupting the conduction of vibrations from the tympanic membrane to the cochlea. This causes progressive unilateral hearing loss, which may advance to bilateral deafness. Other symptoms include tinnitus and paracusis of Willis (hearing conversation better in a noisy environment than in a quiet one).

## **Diagnosis**

Early diagnosis is based on a Rinne test that shows bone conduction lasting longer than air conduction (normally, the reverse is true). As otosclerosis progresses, bone conduction also deteriorates. Audiometric testing reveals hearing loss ranging from 60 dB in early stages to total loss. Weber test detects sound lateralizing to the more affected ear. Physical examination

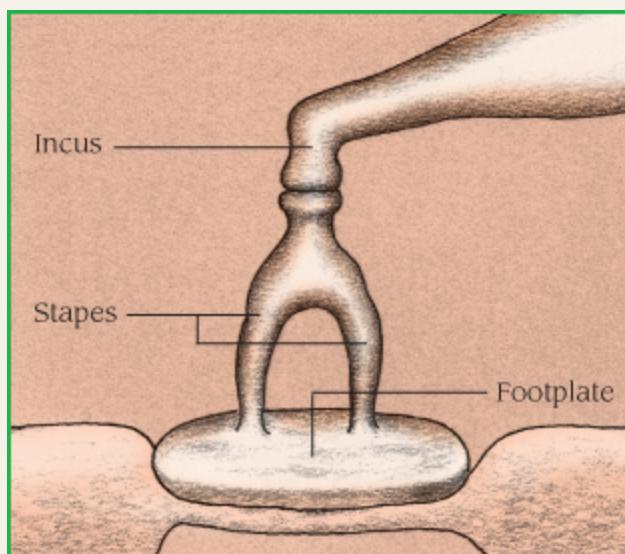
reveals a normal tympanic membrane. Head CT scan and X-ray help distinguish otosclerosis from other causes of hearing loss.

## Treatment

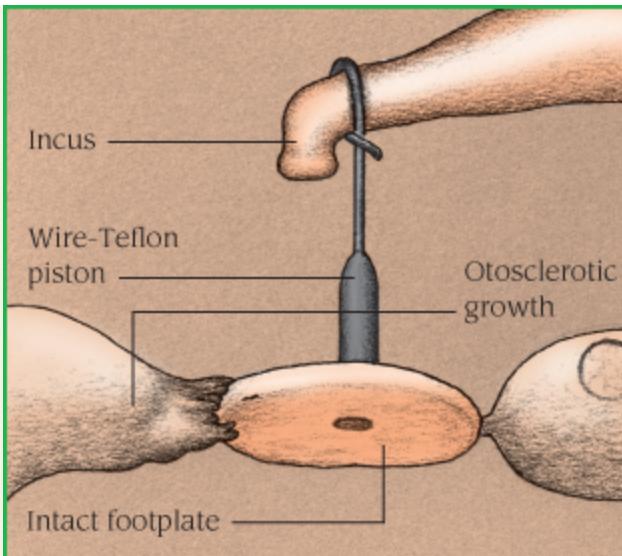
Treatment consists of stapedectomy (removal of the stapes) and insertion of a prosthesis to restore partial or total hearing. This procedure is performed on only one ear at a time, beginning with the ear that has suffered greater damage. Alternative surgery includes stapedotomy (creation of a small hole in the stapes' footplate), through which a wire and piston are inserted. (See *Types of stapedectomy*.) Recent procedural innovations involve laser surgery. Postoperatively, treatment includes antibiotics to prevent infection. If surgery isn't possible, a hearing aid (air conduction aid with molded ear insert receiver) enables the patient to hear conversation in normal surroundings, although this therapy isn't as effective as stapedectomy.

## Types of Stapedectomy

Surgery may remove part or all of the stapes, depending on the extent of otosclerotic growth. It may be performed using various techniques. Two techniques used to implant prostheses are depicted below.

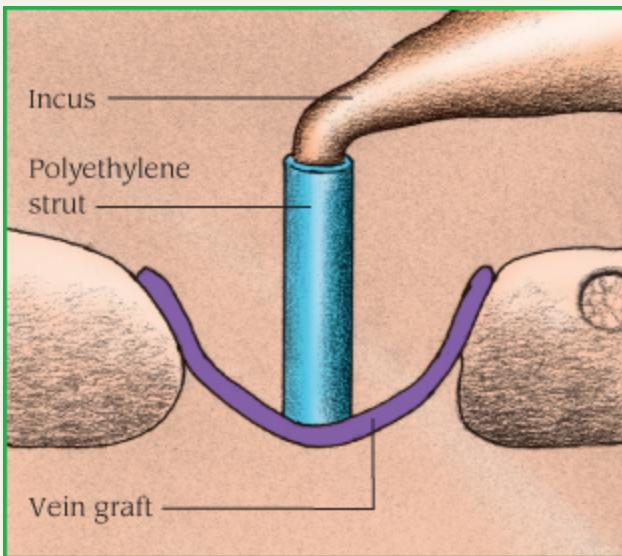


Normal Middle Ear



Partial Stapedectomy

### Wire-Teflon Prosthesis



Total Stapedectomy

### Vein Graft and Strut Prosthesis

## Special Considerations

- During the first 24 hours after surgery, keep the patient supine, with the affected ear facing upward (to maintain the position of the graft). Enforce bed rest with bathroom privileges for 48 hours. Because the

patient may be dizzy, keep the side rails up, and assist them with ambulation. Assess for pain and vertigo, which may be relieved with repositioning or prescribed medication.

- ◆ Tell the patient that hearing won't return until edema subsides and packing is removed.



**ALERT** *Watch for and report postoperative facial drooping, which may indicate swelling of or around the facial nerve.*

- ◆ Before discharge, instruct the patient to avoid loud noises and sudden pressure changes (such as those that occur while diving or flying) until healing is complete (usually 6 months). Advise the patient not to blow their nose for at least 1 week to prevent contaminated air and bacteria from entering the eustachian tube.
- ◆ Stress the importance of protecting the ears against cold; avoiding any activities that provoke dizziness, such as straining, bending, or heavy lifting and, if possible, avoiding contact with anyone who has an URTI. Teach the patient and family how to change the external ear dressing (eye or gauze pad) and care for the incision. Emphasize the need to complete the prescribed antibiotic regimen and to return for scheduled follow-up care.

## INFECTIOUS MYRINGITIS

Acute infectious myringitis is characterized by inflammation, hemorrhage, and effusion of fluid into the tissue at the end of the external ear canal and the tympanic membrane. This self-limiting disorder (resolving spontaneously within 3 days to 2 weeks) commonly follows acute otitis media or URTI.

Chronic granular myringitis, a rare inflammation of the squamous layer of the tympanic membrane, causes gradual hearing loss. Without specific treatment, this condition can lead to stenosis of the ear canal, as granulation extends from the tympanic membrane to the external ear.

### Causes and Incidence

Acute infectious myringitis usually follows viral infection but may also result from infection with bacteria (pneumococcus, *H. influenzae*, beta-hemolytic streptococci, staphylococci) or any other organism that can cause

acute otitis media. Myringitis is a rare sequela of atypical pneumonia caused by *Mycoplasma pneumoniae*. The cause of chronic granular myringitis is unknown.

Acute infectious myringitis frequently occurs epidemically in children.

## Pathophysiology

Bullous myringitis is a common condition characterized by vesicular eruptions of the tympanic membrane. In the majority of cases the condition is self-limited, although serious complications have been reported. The disease is primarily one of childhood, but is frequently seen in adults. Bullous myringitis is generally thought to be of viral origin, although several investigations have failed to establish this. Recent studies suggest a relationship to influenza virus and the Eaton agent, a pleuropneumonia-like organism (*M. pneumoniae*) known to be capable of producing primary atypical pneumonia.

## Complications

- ◆ Gradual hearing loss
- ◆ Stenosis of the ear canal

## Signs and Symptoms

Acute infectious myringitis begins with severe ear pain, commonly accompanied by tenderness over the mastoid process. Small, reddened, inflamed blebs form in the canal, on the tympanic membrane, and, with bacterial invasion, in the middle ear. Fever and hearing loss are rare unless fluid accumulates in the middle ear or a large bleb totally obstructs the external auditory meatus. Spontaneous rupture of these blebs may cause bloody discharge. Chronic granular myringitis produces pruritus, purulent discharge, and gradual hearing loss.

## Diagnosis

 **CONFIRMING DIAGNOSIS** *Diagnosis of acute infectious myringitis is based on physical examination showing characteristic blebs and a typical patient history. Culture and sensitivity testing of exudate identifies secondary infection. In chronic granular myringitis, physical examination*

*may reveal granulation extending from the tympanic membrane to the external ear.*

## Treatment

Hospitalization usually isn't required for acute infectious myringitis. Treatment consists of measures to relieve pain: analgesics, such as aspirin or acetaminophen, and application of heat to the external ear are usually sufficient, but severe pain may necessitate the use of codeine.



**ALERT** *Aspirin and combination aspirin products aren't recommended for people younger than age 19 during episodes of fever-causing illnesses because the use of aspirin has been linked to Reye syndrome.*

Systemic or topical antibiotics prevent or treat secondary infection. Incision of blebs and evacuation of serum and blood may relieve pressure and help drain exudate but don't speed recovery.

Treatment for chronic granular myringitis consists of systemic antibiotics or local anti-inflammatory/antibiotic combination eardrops, and surgical excision and cautery. If stenosis is present, surgical reconstruction is necessary.

## Special Considerations

- ◆ Stress the importance of completing the prescribed antibiotic therapy.
- ◆ Teach the patient how to instill topical antibiotics (eardrops). When necessary, explain incision of blebs.



**PREVENTION** *Advise early treatment for acute otitis media.*

## Inner Ear

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### MÉNIÈRE DISEASE

Ménière disease, a labyrinthine dysfunction also known as *endolymphatic hydrops*, produces severe vertigo, sensorineural hearing loss, and tinnitus. After multiple attacks over several years, this disorder leads to residual tinnitus and hearing loss. Usually, only one ear is involved.

## **Causes and Incidence**

The exact cause of Ménière disease is unknown. It may result from overproduction or decreased absorption of endolymph, which causes endolymphatic hydrops or endolymphatic hypertension, with consequent degeneration of the vestibular and cochlear hair cells. This condition may also stem from autonomic nervous system dysfunction that produces a temporary constriction of blood vessels supplying the inner ear. In some cases, Ménière disease may be related to otitis media, syphilis, or head injury. Risk factors include recent viral illness, respiratory infection, stress, fatigue, use of prescription or nonprescription drugs (such as aspirin), and a history of allergies, smoking, and alcohol use. There also may be genetic risk factors: In some women, premenstrual edema may precipitate attacks of Ménière disease.

In the United States, about 100,000 people per year develop Ménière disease.

## **Pathophysiology**

The pathophysiology of Ménière disease is not clearly understood. It was previously thought that Ménière was closely correlated with endolymphatic hydrops, a condition in which endolymph builds up because of an obstruction in the endolymphatic sac. Other possible origins of the disease are perisaccular fibrosis, atrophy of the endolymphatic sac and loss of epithelial integrity, hypoplasia of the vestibular aqueduct, and narrowing of the lumen of the endolymphatic duct.

## **Complications**

- ◆ Tinnitus
- ◆ Partial to total hearing loss
- ◆ Permanent balance disability

## **Signs and Symptoms**

Ménière disease produces three characteristic effects: severe episodic vertigo, tinnitus, and sensorineural hearing loss. A feeling of fullness or blockage in the ear is also common. Violent paroxysmal attacks last from 10 minutes to several hours. During an acute attack, other symptoms include severe nausea, vomiting, sweating, giddiness, and nystagmus.

Vertigo may cause loss of balance and falling to the affected side. Symptoms tend to wax and wane as the endolymphatic pressure rises and falls. To lessen these symptoms, the patient may assume a characteristic posture—lying on the side of the unaffected ear and looking in the direction of the affected ear.

Initially, the patient may be asymptomatic between attacks, except for residual tinnitus that worsens during an attack. Such attacks may occur several times a year, or remissions may last as long as several years. These attacks become less frequent as hearing loss progresses (usually unilaterally); they may cease when hearing loss is total. All symptoms are aggravated by motion.

## Diagnosis

The presence of all three typical symptoms suggests Ménière disease. Audiometric studies indicate a sensorineural hearing loss and loss of discrimination and recruitment. Selected studies such as electronystagmography, electrocochleography, CT scan, magnetic resonance imaging, or X-rays of the internal meatus may be necessary for differential diagnosis.

Laboratory studies, including thyroid and lipid studies, may be performed to rule out other conditions such as *Treponema pallidum*.

Caloric testing may reveal loss or impairment of thermally induced nystagmus on the involved side. However, it's important not to overlook an acoustic tumor, which produces an identical clinical picture.

## Treatment

Treatment with atropine may stop an attack in 20 to 30 minutes. Epinephrine or diphenhydramine may be necessary in a severe attack; dimenhydrinate, meclizine, diphenhydramine, or diazepam may be effective in a milder attack.

Long-term management includes use of a diuretic or vasodilator and restricted sodium intake (<2 g/day). A typical diuretic regime is hydrochlorothiazide 50 to 100 mg daily. Prophylactic antihistamines or mild sedatives (phenobarbital, diazepam) may also be helpful. If Ménière disease persists after 2 years of treatment, produces incapacitating vertigo, or resists medical management, surgery may be necessary. Destruction of the affected labyrinth permanently relieves symptoms but results in

irreversible hearing loss. Systemic streptomycin is reserved for the patient with bilateral disease for whom no other treatment can be considered. If a patient fails medical therapy and remains disabled by vertigo, surgical decompression of the endolymphatic sac may bring relief.

## **Special Considerations**

If the patient is in the hospital during an attack of Ménière disease:

- ◆ Advise the patient against reading and exposure to glaring lights, to reduce dizziness.
- ◆ Keep the side rails of the patient's bed up to prevent falls. Tell the patient not to get out of bed or walk without assistance.
- ◆ Instruct the patient to avoid sudden position changes and any tasks that vertigo makes hazardous because an attack can begin quite rapidly. Hazardous activities, such as driving and climbing, should be avoided until 1 week after symptoms disappear.
- ◆ Before surgery, if the patient is vomiting, record fluid intake and output and characteristics of vomitus. Administer antiemetics as needed, and give small amounts of fluid frequently.
- ◆ After surgery, record intake and output carefully. Tell the patient to expect dizziness and nausea for 1 or 2 days after surgery. Give prophylactic antibiotics and antiemetics, as ordered.

## **LABYRINTHITIS**

Labyrinthitis, an inflammation of the labyrinth of the inner ear, frequently incapacitates the patient by producing severe vertigo that lasts for 3 to 5 days; symptoms gradually subside over a 3- to 6-week period. Viral labyrinthitis is commonly associated with URTI.

## **Causes**

Labyrinthitis is usually caused by viral infection. It may be a primary infection, the result of trauma, or a complication of influenza, otitis media, or meningitis. In chronic otitis media, cholesteatoma formation erodes the bone of the labyrinth, allowing bacteria to enter from the middle ear. Toxic drug ingestion is another possible cause of labyrinthitis and neuritis.

## **Pathophysiology**

Labyrinthitis is an inflammatory response within the membranous inner ear structures in response to infection. It is a generally short-lived minor illness that has the potential to cause temporary or permanent disablement in terms of hearing loss. Other symptoms include nausea and vomiting, pain in the affected ear, vertigo, and fever.

## Complications

- ◆ Meningitis
- ◆ Permanent hearing loss
- ◆ Permanent balance disability

## Signs and Symptoms

Because the inner ear controls both hearing and balance, this infection typically produces severe vertigo (with any movement of the head) and sensorineural hearing loss. Vertigo begins gradually but peaks within 48 hours, causing loss of balance and falling in the direction of the affected ear. Other associated signs and symptoms include spontaneous nystagmus, with jerking movements of the eyes toward the unaffected ear, and nausea, vomiting, and giddiness. With cholesteatoma, signs of middle ear disease may appear. With severe bacterial infection, purulent drainage, increased salivation, generalized malaise, and perspiration can occur. To minimize symptoms such as giddiness and nystagmus, the patient may assume a characteristic posture—lying on the side of the unaffected ear and looking in the direction of the affected ear.

## Diagnosis

A typical clinical picture and a history of URTI suggest labyrinthitis. Typical diagnostic measures include culture and sensitivity testing to identify the infecting organism, if purulent drainage is present, and audiometric testing. When an infectious etiology can't be found, additional testing must be done to rule out a brain lesion or Ménière disease.

Differentiation from other causes of dizziness or vertigo may include head CT scan or magnetic resonance imaging, audiology or audiometry testing, caloric stimulation tests, electronystagmography, electroencephalogram, and auditory-evoked potential studies.

## Treatment

Symptomatic treatment includes bed rest, with the head immobilized between pillows, and antibiotics to combat diffuse purulent labyrinthitis. Oral fluids can prevent dehydration caused by vomiting. For severe nausea and vomiting, I.V. fluids may be necessary. Medications that help reduce symptoms include antihistamines, anticholinergics, sedative-hypnotics, and antiemetics; benzodiazepines help control vertigo.

When conservative management fails, treatment necessitates surgical excision of the cholesteatoma and drainage of the infected areas of the middle and inner ear. Prevention is possible by early and vigorous treatment for predisposing conditions, such as otitis media and any local or systemic infection.

## Special Considerations

- ◆ Keep the side rails up to prevent falls. Tell the patient to keep still and rest during attacks and to avoid sudden position changes.
- ◆ If vomiting is severe, administer antiemetics as ordered. Record intake and output, and give I.V. fluids as ordered.
- ◆ During an attack, dim the lighting and tell the patient to avoid reading.
- ◆ Tell the patient that recovery may take as long as 6 weeks. During this time, they should limit activities that vertigo may make hazardous. Hazardous activities, such as driving and climbing, should be avoided until 1 week after symptoms disappear.
- ◆ If recovery doesn't occur within 4 to 6 weeks, a CT scan should be performed to rule out an intracranial lesion.

## HEARING LOSS

Hearing loss results from a mechanical or nervous impediment to the transmission of sound waves. The major forms of hearing loss are classified as *conductive loss* (interrupted passage of sound from the external ear to the junction of the stapes and oval window), *sensorineural loss* (impaired cochlea or acoustic [eighth cranial] nerve dysfunction, causing failure of transmission of sound impulses within the inner ear or brain), or *mixed loss* (combined dysfunction of conduction and sensorineural transmission). Hearing loss may be partial or total and is calculated from this American Medical Association formula: Hearing is 1.5% impaired for every decibel that the pure tone average exceeds 25 dB.

## **Causes and Incidence**

*Congenital hearing loss* may be transmitted as a dominant, autosomal dominant, autosomal recessive, or sex-linked recessive trait. Hearing loss in neonates may also result from trauma, toxicity, or infection during pregnancy or delivery. Predisposing factors include a family history of hearing loss or known hereditary disorders (e.g., otosclerosis), maternal exposure to rubella or syphilis during pregnancy, use of ototoxic drugs during pregnancy, prolonged fetal anoxia during delivery, and congenital abnormalities of the ears, nose, or throat. Premature or low-birth-weight neonates are most likely to have structural or functional hearing impairment; those with serum bilirubin levels above 20 mg/dL also risk hearing impairment from the toxic effect of high-serum bilirubin levels on the brain. In addition, trauma during delivery may cause intracranial hemorrhage and may damage the cochlea or the acoustic nerve.

*Sudden deafness* refers to sudden hearing loss in a person with no prior hearing impairment. This condition is considered a medical emergency because prompt treatment may restore full hearing. Its causes and predisposing factors may include:

- ◆ acute infections, especially mumps (most common cause of unilateral sensorineural hearing loss in children), and other bacterial and viral infections, such as rubella, rubeola, influenza, herpes zoster, and infectious mononucleosis; and mycoplasma infections
- ◆ blood dyscrasias (leukemia, hypercoagulation)
- ◆ head trauma or brain tumors
- ◆ metabolic disorders (diabetes mellitus, hypothyroidism, hyperlipoproteinemia)
- ◆ neurologic disorders (multiple sclerosis, neurosyphilis)
- ◆ ototoxic drugs (tobramycin, streptomycin, quinine, gentamicin, furosemide, ethacrynic acid)
- ◆ vascular disorders (hypertension, arteriosclerosis)

*Noise-induced hearing loss*, which may be transient or permanent, may follow prolonged exposure to loud noise (85 to 90 dB) or brief exposure to extremely loud noise (>90 dB). Such hearing loss is common in workers subjected to constant industrial noise and in military personnel, hunters, and rock musicians.

*Presbycusis*, an otologic effect of aging, results from a loss of hair cells in the organ of Corti. This disorder causes progressive, symmetrical, bilateral sensorineural hearing loss, usually of high-frequency tones.

Minor decreases in hearing are common after age 20. Some deafness due to nerve damage occurs in one of every five people by age 55.

## Complications

- ◆ Tympanic membrane perforation
- ◆ Cholesteatoma
- ◆ Permanent hearing loss

## Signs and Symptoms



**PEDIATRIC TIP** Although congenital hearing loss may produce no obvious signs of hearing impairment at birth, a deficient response to auditory stimuli generally becomes apparent within 2 to 3 days. As the child grows older, hearing loss impairs speech development.

Sudden deafness may be conductive, sensorineural, or mixed, depending on etiology. Associated clinical features depend on the underlying cause.

Noise-induced hearing loss causes sensorineural damage, the extent of which depends on the duration and intensity of the noise. Initially, the patient loses perception of certain frequencies (around 4,000 Hz) but, with continued exposure, eventually loses perception of all frequencies.



**ELDER TIP** Presbycusis usually produces tinnitus and the inability to understand the spoken word.



**PEDIATRIC TIP** The behavior of an infant who's deaf may appear normal and mislead the parents as well as the professional, especially if the infant has autosomal recessive deafness and is the first child of carrier parents.

## Diagnosis



**CONFIRMING DIAGNOSIS** Patient, family, and occupational histories and a complete audiology examination usually provide ample evidence of

*hearing loss and suggest possible causes or predisposing factors.*

The Weber, Rinne, and specialized audiology tests differentiate between conductive and sensorineural hearing loss.

## **Treatment**

After the underlying cause is identified, therapy for congenital hearing loss refractory to surgery consists of developing the patient's ability to communicate through sign language, speech reading, or other effective means. Measures to prevent congenital hearing loss include aggressively immunizing children against rubella to reduce the risk of maternal exposure during pregnancy; educating pregnant women about the dangers of exposure to drugs, chemicals, or infection; and careful monitoring during labor and delivery to prevent fetal anoxia.

Treatment for sudden deafness requires prompt identification of the underlying cause. Prevention necessitates educating patients and healthcare professionals about the many causes of sudden deafness and the ways to recognize and treat them.

Hyperbilirubinemia can be controlled by phototherapy and exchange transfusions. Children need the appropriate immunizations. Medications that may be ototoxic should be used judiciously in children and monitored closely. Reduction of exposure to loud noises generally prevents high-frequency hearing loss.

In people with noise-induced hearing loss, overnight rest usually restores normal hearing in those who have been exposed to noise levels greater than 90 dB for several hours, but not in those who have been exposed to such noise repeatedly. As hearing deteriorates, treatment must include speech and hearing rehabilitation, because hearing aids are seldom helpful. Prevention of noise-induced hearing loss requires public recognition of the dangers of noise exposure and insistence on the use, as mandated by law, of protective devices such as earplugs during occupational exposure to noise.

Amplifying sound, as with a hearing aid, helps some patients with presbycusis, but many patients have an intolerance to loud noise and wouldn't be helped by a hearing aid.

## **Special Considerations**

- ◆ When speaking to a patient with hearing loss who can read lips, stand directly in front of them, with the light on your face, and speak slowly and distinctly. If possible, speak to the patient at eye level. Approach the patient within their visual range, and elicit their attention by raising your arm or waving; touching them may be unnecessarily startling.
- ◆ Make other staff members and facility personnel aware of the patient's disability and their established method of communication. Carefully explain diagnostic tests and facility procedures in a way the patient understands.
- ◆ Make sure the patient with a hearing loss is in an area where activity can be observed and approaching persons can be seen because such a patient depends totally on visual clues.
- ◆ When addressing an older patient, speak slowly and distinctly in a low tone; avoid shouting.
- ◆ Provide emotional support and encouragement to the patient learning to use a hearing aid. Teach them how the aid works and how to maintain it.
- ◆ Refer children with suspected hearing loss to an audiologist or otolaryngologist for further evaluation. Any child who fails a language screening examination should be referred to a speech pathologist for language evaluation. The child with a mild language delay may be involved with a home language-enrichment program.



**PREVENTION** *Watch for signs of hearing impairment in the patient receiving ototoxic drugs. Emphasize the danger of excessive exposure to noise; stress the danger to pregnant women of exposure to drugs, chemicals, and infection (especially rubella); and encourage the use of protective devices in a noisy environment.*

## MOTION SICKNESS

Motion sickness is characterized by loss of equilibrium associated with nausea and vomiting that results from irregular or rhythmic movements or from the sensation of motion. Removal of the stimulus restores normal equilibrium. Motion sickness also can be induced when patterns of motion differ from what the patient has previously experienced.

### Causes and Incidence

Motion sickness may result from excessive stimulation of the labyrinthine receptors of the inner ear by certain motions, such as those experienced in a car, boat, plane, or swing. The disorder may also be caused by confusion in the cerebellum from conflicting sensory input—the visual stimulus (a moving horizon) conflicts with labyrinthine perception. Predisposing factors include tension or fear, offensive odors, or sights and sounds associated with a previous attack. Motion sickness from cars, elevators, trains, and swings is most common in children; from boats and airplanes, in adults. People who suffer from one kind of motion sickness aren't necessarily susceptible to other types.

## **Pathophysiology**

Motion sickness is a syndrome that occurs when a patient is exposed to certain types of motion and usually resolves soon after its cessation. It is a common response to motion stimuli during travel. Although nausea is a hallmark symptom, the syndrome includes symptoms ranging from vague malaise to completely incapacitating illness. These symptoms, which can affect the patient's recreation, employment, and personal safety, can occur within minutes of experiencing motion and can last for several hours after its cessation.

## **Signs and Symptoms**

Typically, motion sickness induces nausea, vomiting, headache, dizziness, fatigue, diaphoresis, and, occasionally, difficulty in breathing, leading to a sensation of suffocation. These symptoms usually subside when the precipitating stimulus is removed, but they may persist for several hours or days.

## **Treatment**

The best way to treat the disorder is to stop the motion that's causing it. If this isn't possible, the patient will benefit from lying down, closing their eyes, and trying to sleep. Antiemetics, such as dimenhydrinate, cyclizine, meclizine, and scopolamine (transdermal patch), may prevent or relieve motion sickness.

## **Special Considerations**



**PEDIATRIC TIP** *An elevated car seat may help prevent motion sickness in a child by allowing the patient to see out of the front window.*

- ◆ Tell the patient to avoid exposure to precipitating motion whenever possible.
- ◆ Instruct the patient to avoid eating or drinking for at least 4 hours before traveling and to take an antiemetic 30 to 60 minutes before traveling or to apply a transdermal scopolamine patch at least 4 hours before traveling. Tell the patient with prostate enlargement or glaucoma to consult a physician or pharmacist before taking antiemetics.



**PREVENTION** *The traveler can minimize motion sickness by sitting where motion is least apparent (near the wing section in an aircraft, in the center of a boat, or in the front seat of an automobile). Instruct the patient to keep the head still and eyes closed or focused on a distant and stationary object.*

## Nose

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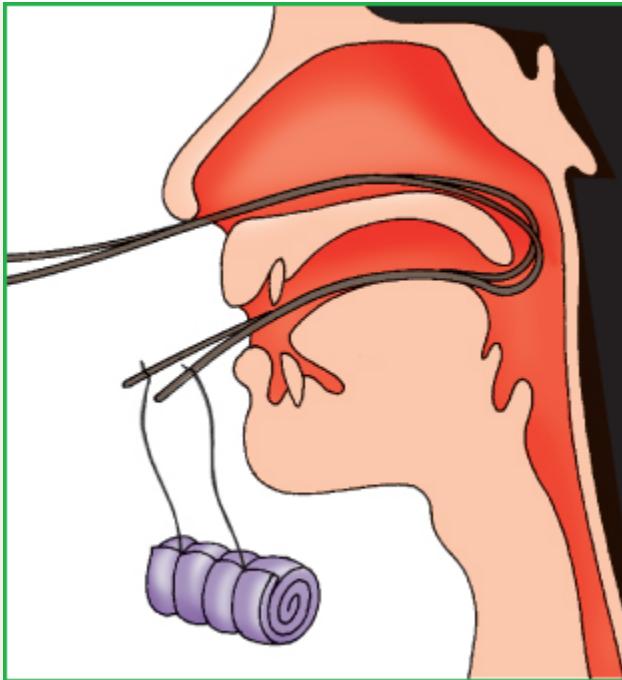
### EPISTAXIS

Epistaxis, commonly known as a *nosebleed*, may be a primary disorder or may occur secondary to another condition. Such bleeding in children generally originates in the anterior nasal septum and tends to be mild. In adults, such bleeding is most likely to originate in the posterior septum and can be severe enough to warrant nasal packing. (See *Inserting an anterior–posterior nasal pack*, pages 644 and 645.) Epistaxis is twice as common in children as in adults.

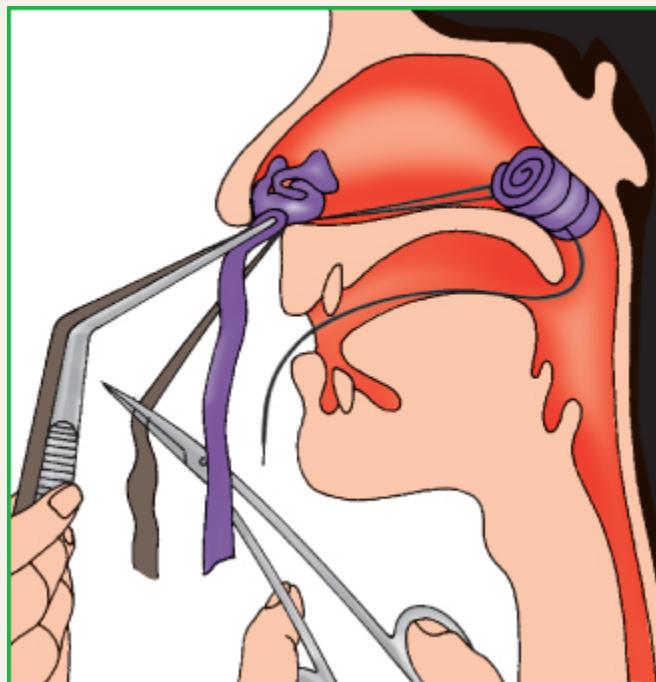
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### Inserting an Anterior–Posterior Nasal Pack

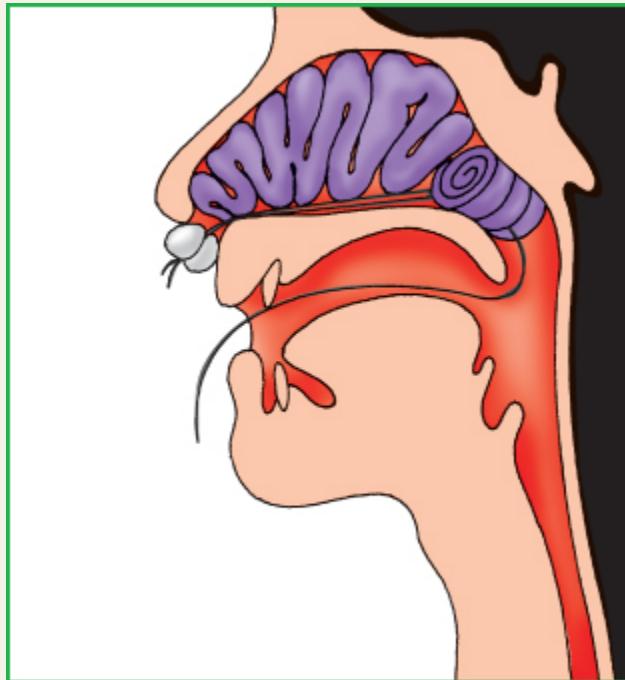
The first step in the insertion of an anterior–posterior nasal pack is the insertion of catheters into the nostrils. After the catheters are drawn through the mouth, a suture from the pack is tied to each (as shown above).



This positions the pack in place as the catheters are drawn back through the nostrils. Although the sutures are held tightly, packing is inserted into the anterior nose (as shown above).



The sutures are then secured around a dental roll; the middle suture extends from the mouth (as shown above) and is taped to the cheek.



## Causes

Epistaxis usually follows trauma from external or internal causes: a blow to the nose, nose picking, or insertion of a foreign body; low humidity; or allergies, colds, or sinusitis. Less commonly, it follows polyps; acute or chronic infections such as sinusitis or rhinitis, which cause congestion and eventual bleeding of the capillary blood vessels; or inhalation of chemicals that irritate the nasal mucosa.

Predisposing factors include anticoagulant therapy, hypertension, long-term use of aspirin, overuse of decongestant nasal sprays, high altitudes and dry climates, sclerotic vessel disease, Hodgkin disease, hereditary hemorrhagic telangiectasia, neoplastic disorders (such as juvenile nasopharyngeal angiofibromas [JNAs]), scurvy, vitamin K deficiency, rheumatic fever, and blood dyscrasias (hemophilia, purpura, leukemia, and anemias).

## Pathophysiology

Nosebleeds are due to the rupture of a blood vessel within the richly perfused nasal mucosa. Rupture may be spontaneous or initiated by trauma. An increase in blood pressure (e.g., due to general hypertension) tends to increase the duration of spontaneous epistaxis. Anticoagulant medication and disorders of blood clotting can promote and prolong bleeding. Spontaneous epistaxis is more common in the elderly as the nasal mucosa (lining) becomes dry and thin and blood pressure tends to be higher. The elderly are also more prone to prolonged nose bleeds as their blood vessels are less able to constrict and control the bleeding. Sometimes blood flowing from other sources of bleeding passes through the nasal cavity and exits the nostrils. It is thus blood coming from the nose but is not a true nosebleed, that is, not truly originating from the nasal cavity.

## Complications

- ◆ Aspiration
- ◆ Shock

## Signs and Symptoms

Blood oozing from the nostrils usually originates in the anterior nose and is bright red. Blood from the back of the throat originates in the posterior area and may be dark or bright red (commonly mistaken for hemoptysis due to expectoration). Epistaxis is generally unilateral, except when it's because of dyscrasia or severe trauma. In severe epistaxis, blood may seep behind the nasal septum; it may also appear in the middle ear and in the corners of the eyes.

Associated clinical effects depend on the severity of bleeding. Moderate blood loss may produce light-headedness, dizziness, and slight respiratory difficulty; severe hemorrhage causes hypotension, rapid and bounding pulse, dyspnea, and pallor. Bleeding is considered severe if it persists longer than 10 minutes after pressure is applied and causes blood loss as great as 1 L/hour in adults. Exsanguination (bleeding to death) from epistaxis is rare.

## Diagnosis



**CONFIRMING DIAGNOSIS** *Although simple observation confirms epistaxis, inspection with a bright light and a nasal speculum is necessary to locate the site of bleeding.*

Relevant laboratory values include:

- ◆ gradual reduction in hemoglobin levels and hematocrit (HCT; usually inaccurate immediately following epistaxis because of hemoconcentration)
- ◆ decreased platelet count in the patient with blood dyscrasia
- ◆ prothrombin time and partial thromboplastin time showing a coagulation time twice the control, because of a bleeding disorder or anticoagulant therapy

Diagnosis must rule out underlying systemic causes of epistaxis, especially disseminated intravascular coagulation and rheumatic fever. Bruises or concomitant bleeding elsewhere probably indicates a hematologic disorder.



**PEDIATRIC TIP** *Bleeding tests are indicated if any of the following are present:*

- ◆ *family history of a bleeding disorder*
- ◆ *medical history of easy bleeding*
- ◆ *spontaneous bleeding at other sites*
- ◆ *onset before age 2 or a drop in HCT due to epistaxis*
- ◆ *bleeding that won't clot with direct pressure by the physician*
- ◆ *bleeding that lasts longer than 30 minutes*

## Treatment

Mild nosebleeds that occur spontaneously may be treated by gently squeezing the soft portion of the nose between the thumb and finger for 5 to 10 minutes while the patient leans forward slightly (to avoid swallowing the blood) and breathes through the mouth.

For anterior bleeding, treatment consists of application to the bleeding site of a cotton ball saturated with epinephrine, and external pressure, followed by cauterization with electrocautery or a silver nitrate stick. If these measures don't control the bleeding, petroleum gauze nasal packing may be needed.

For posterior bleeding, therapy includes gauze packing inserted through the nose, or postnasal packing inserted through the mouth, depending on the

bleeding site. (Gauze packing generally remains in place for 24 to 48 hours; postnasal packing, 3 to 5 days.) An alternate method, the nasal balloon catheter, also controls bleeding effectively. Antibiotics may be appropriate if packing must remain in place for longer than 24 hours. If local measures fail to control bleeding, additional treatment may include supplemental vitamin K and, for severe bleeding, blood transfusions and surgical ligation or embolization of a bleeding artery.

## Special Considerations

To control epistaxis:

- ◆ Elevate the patient's head to 45 degrees.
- ◆ Continuously compress the soft portion of the nares against the septum for 5 to 10 minutes. Apply an ice collar or cold, wet compresses to the nose. If bleeding continues after 10 minutes of pressure, notify the physician.
- ◆ Administer oxygen as needed, and monitor saturation levels.
- ◆ Monitor vital signs and skin color; record blood loss.
- ◆ Tell the patient to breathe through their mouth and not to swallow blood, talk, or blow their nose.
- ◆ Keep vasoconstrictors, such as phenylephrine, handy.
- ◆ Reassure the patient and their family that epistaxis usually looks worse than it is.



### PREVENTION

- ◆ *Instruct the patient not to pick their nose or insert foreign objects into it, and to avoid bending or lifting. Emphasize the need for follow-up examinations and periodic blood studies after an episode of epistaxis. Advise prompt treatment for nasal infection or irritation.*
- ◆ *Suggest humidifiers for people who live in dry climates or at high elevations, or whose homes are heated with circulating hot air.*

## SEPTAL PERFORATION AND DEVIATION

Perforated septum, a hole in the nasal septum between the two air passages, usually occurs in the anterior cartilaginous septum but may occur in the bony septum. Deviated septum, a shift from the midline, is common in most

adults. This condition may be severe enough to obstruct the passage of air through the nostrils. With surgical correction, the prognosis for either perforated or deviated septum is good.

## **Causes and Incidence**

Generally, perforated septum is caused by traumatic irritation, most commonly resulting from excessive nose picking; less frequently, it results from repeated cauterization for epistaxis or from penetrating septal injury. It may also result from perichondritis, an infection that gradually erodes the perichondrial layer and cartilage, finally forming an ulcer that perforates the septum. Other causes of septal perforation include syphilis, tuberculosis, untreated septal hematoma, inhalation of irritating chemicals, cocaine snorting, use of nasal sprays, chronic nasal infections, nasal carcinoma, granuloma, and chronic sinusitis.

Deviated septum commonly develops during normal growth, as the septum shifts from one side to the other. Consequently, few adults have perfectly straight septa. Nasal trauma resulting from a fall, a blow to the nose, or surgery further exaggerates the deviation. Congenital deviated septum is rare.

## **Complications**

- ◆ Hemorrhage
- ◆ Infections
- ◆ Deformity

## **Signs and Symptoms**

A small septal perforation is usually asymptomatic but may produce a whistle on inspiration. A large perforation causes rhinitis, epistaxis, nasal crusting, and watery discharge.

The patient with a deviated septum may develop a crooked nose, as the midline deflects to one side. The predominant symptom of severe deflection, however, is nasal obstruction. Other manifestations include a sensation of fullness in the face, shortness of breath, stertor (snoring or laborious breathing), nasal discharge, recurring epistaxis, infection, sinusitis, and headache.

## **Diagnosis**

Although clinical features suggest septal perforation or deviation, confirmation requires inspection of the nasal mucosa with a bright light and a nasal speculum.

## Treatment

Symptomatic treatment for perforated septum includes decongestants to reduce nasal congestion by local vasoconstriction, local application of lanolin or petroleum jelly to prevent ulceration and crusting, and antibiotics to combat infection. Surgery may be necessary to graft part of the perichondrial layer over the perforation. Also, a plastic or Silastic “button” prosthesis may be used to close the perforation.

Symptomatic treatment for deviated septum usually includes analgesics to relieve headache, decongestants to minimize secretions, and, as necessary, vasoconstrictors, nasal packing, or cautery to control hemorrhage. Manipulation of the nasal septum at birth can correct congenital deviated septum.

Corrective surgical procedures include:

- ◆ reconstruction of the nasal septum by submucous resection to reposition the nasal septal cartilage and relieve nasal obstruction
- ◆ rhinoplasty to correct nasal structure deformity by intranasal incisions
- ◆ septoplasty to relieve nasal obstruction and enhance cosmetic appearance

## Special Considerations

- ◆ In the patient with perforated septum, use a cotton applicator to apply petroleum jelly to the nasal mucosa to minimize crusting and ulceration.
- ◆ Warn the patient with perforation or severe deviation against blowing their nose. To relieve nasal congestion, instill saline nose drops and suggest use of a humidifier. Give decongestants as ordered.
- ◆ Prevention and patient education are the first lines of treatment for perforations caused by nasal sprays. Proper technique (aiming away from the nasal septum) should be reviewed. Medication should be withheld when scabs are noted on the septum.
- ◆ To treat epistaxis, have the patient sit upright, provide an emesis basin, and instruct the patient to expectorate any blood. Compress the outer portion of the nose against the septum for 10 to 15 minutes, and apply ice packs. If bleeding persists, notify the physician.

- ◆ If corrective surgery is scheduled, prepare the patient to expect postoperative facial edema, periorbital bruising, and nasal packing, which remains in place for 12 to 24 hours. The patient must breathe through the mouth. After surgery for deviated septum, the patient may also have a splint on their nose.
- ◆ To reduce or prevent edema and promote drainage, place the patient in semi-Fowler position, and use a cool-mist vaporizer to liquefy secretions and facilitate normal breathing. To lessen facial edema and pain, place crushed ice in a rubber glove or a small ice bag, and apply the glove or ice bag intermittently over the eyes and nose for 24 hours.
- ◆ Because the patient is breathing through the mouth, provide frequent mouth care.
- ◆ Change the mustache dressing or drip pad as needed. Record the color, consistency, and amount of drainage. While nasal packing is in place, expect slight, bright red drainage, with clots. After packing is removed, watch for purulent discharge, an indication of infection.
- ◆ Watch for and report excessive swallowing, hematoma, or a falling or flapping septum (depressed, or soft and unstable septum). Intranasal examination is necessary to detect hematoma formation. Any of these complications requires surgical correction.
- ◆ Administer sedatives and analgesics as needed. Because of its anticoagulant properties, aspirin is contraindicated after surgery for septal deviation or perforation.
- ◆ Nose blowing may cause bruising and swelling even after nasal packing is removed. After surgery, the patient must limit physical activity for 2 or 3 days and, if they are a smoker, they must stop smoking for at least 2 days.
- ◆ Instruct the patient to sneeze with their mouth open and to avoid bending over at the waist. (Advise the patient to stoop to pick up fallen objects.)

## SINUSITIS

Sinusitis—*inflammation of the paranasal sinuses*—may be acute, subacute, chronic, allergic, or hyperplastic. Acute sinusitis usually results from the common cold and lingers in subacute form in only about 10% of patients. Chronic sinusitis follows persistent bacterial infection; allergic sinusitis accompanies allergic rhinitis; hyperplastic sinusitis is a combination of

purulent acute sinusitis and allergic sinusitis or rhinitis. The prognosis is good for all types.

## Causes and Incidence

Sinusitis usually results from viral or bacterial infection. The bacteria responsible for acute sinusitis are usually pneumococci, other streptococci, *H. influenzae*, and *M. catarrhalis*. Staphylococci and gram-negative bacteria are more likely to cause sinusitis in chronic cases or in intensive care patients.

Predisposing factors include any condition that interferes with drainage and ventilation of the sinuses, such as chronic nasal edema, deviated septum, viscous mucus, nasal polyps, allergic rhinitis, nasal intubation, or debilitation due to chemotherapy, malnutrition, diabetes, blood dyscrasias, cystic fibrosis, human immunodeficiency virus or other immunodeficiency disorders, or chronic use of steroids. Bacterial invasion commonly occurs as a result of the conditions listed above or after a viral infection. It may also result from swimming in contaminated water.

Other risk factors for developing sinusitis include a history of asthma, overuse of nasal decongestants, presence of a foreign body in the nose, frequent swimming or diving, dental work, pregnancy, changes in altitude (flying or climbing), air pollution and smoke, gastroesophageal reflux disease (GERD), and having a deviated nasal septum, nasal bone spur, or polyp.

Each year, more than 30 million adults and children get sinusitis.



**PEDIATRIC TIP** *The incidence of both acute and chronic sinusitis increases in later childhood. Sinusitis may be more prevalent in children who have had tonsils and adenoids removed.*

## Complications

- ◆ Meningitis
- ◆ Cavernous and sinus thrombosis
- ◆ Bacteremia or septicemia
- ◆ Brain abscess
- ◆ Osteomyelitis
- ◆ Mucocele

- ◆ Orbital cellulitis abscess

## Pathophysiology

The most common cause of acute sinusitis is an URTI of viral origin. The viral infection can lead to inflammation of the sinuses that usually resolves without treatment in less than 14 days. If symptoms worsen after 3 to 5 days or persist for longer than 10 days and are more severe than normally experienced with a viral infection, a secondary bacterial infection is diagnosed. The inflammation can predispose to the development of acute sinusitis by causing sinus ostial blockage. Although inflammation in any of the sinuses can lead to blockade of the sinus ostia, the most commonly involved sinuses in both acute and chronic sinusitis are the maxillary and the anterior ethmoid sinuses. The nasal mucosa responds to the virus by producing mucus and recruiting mediators of inflammation, such as white blood cells, to the lining of the nose, which cause congestion and swelling of the nasal passages. The resultant sinus cavity hypoxia and mucus retention cause the cilia—which move mucus and debris from the nose—to function less efficiently, creating an environment for bacterial growth. If the acute sinusitis does not resolve, chronic sinusitis can develop from mucus retention, hypoxia, and blockade of the ostia. This promotes mucosal hyperplasia, continued recruitment of inflammatory infiltrates, and the potential development of nasal polyps.

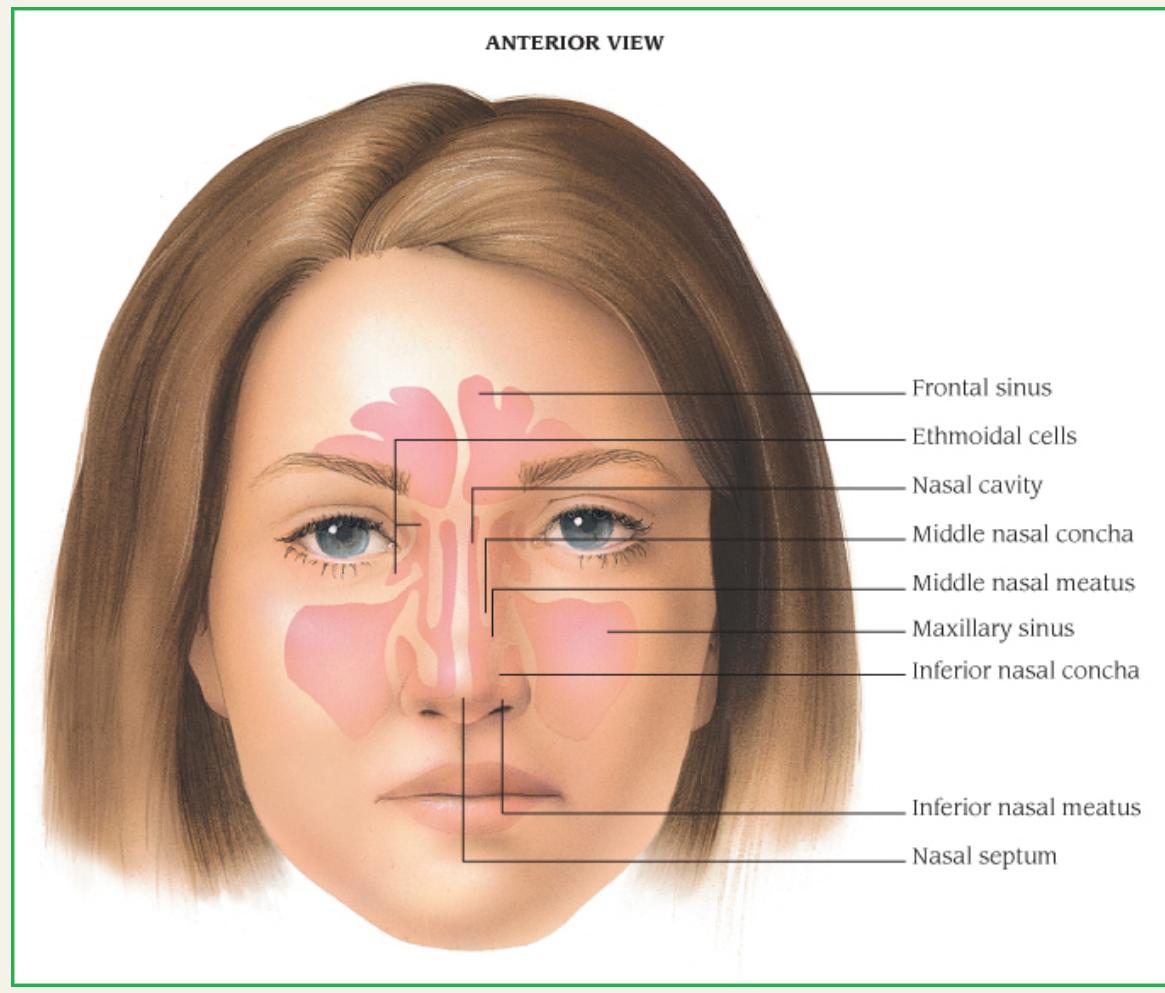
## Signs and Symptoms

The primary indication of acute sinusitis is nasal congestion, followed by a gradual buildup of pressure in the affected sinus. For 24 to 48 hours after onset, nasal discharge may be present and later may become purulent. Associated symptoms include malaise, sore throat, headache, and low-grade fever of 99° to 99.5° F (37.2° to 37.5° C).

Characteristic pain depends on the affected sinus: maxillary sinusitis causes pain over the cheeks and upper teeth; ethmoid sinusitis, pain over the eyes; frontal sinusitis, pain over the eyebrows; and sphenoid sinusitis (rare), pain behind the eyes. (See *Locating the paranasal sinuses*, page 648.)

### Locating The Paranasal Sinuses

The location of a patient's sinusitis pain indicates the affected sinus. For example, an infected maxillary sinus can cause tooth pain. (Note: The sphenoid sinus, which lies under the eye and above the soft palate, isn't depicted here.)



Purulent nasal drainage that continues for longer than 3 weeks after an acute infection subsides suggests *subacute sinusitis*. Other clinical features of the subacute form include nasal congestion, vague facial discomfort, fatigue, and a nonproductive cough.

*Chronic sinusitis* is defined as infection lasting longer than 8 weeks. The effects of chronic sinusitis are similar to those of acute sinusitis, but the chronic form causes continuous mucopurulent discharge.

The effects of *allergic sinusitis* are the same as those of allergic rhinitis. In both conditions, the prominent symptoms are sneezing, frontal headache, watery nasal discharge, and a stuffy, burning, itchy nose.

In *hyperplastic sinusitis*, bacterial growth on the diseased tissue causes pronounced tissue edema; thickening of the mucosal lining and the development of mucosal polyps combine to produce chronic stuffiness of the nose, in addition to headaches.

## Diagnosis

The following measures are useful:

- ◆ Antral puncture promotes drainage of purulent material. It may also be used to provide a specimen for culture and sensitivity testing of the infecting organism, but it's seldom performed.
- ◆ Nasal examination reveals inflammation and pus.
- ◆ Palpation and percussion reveal tenderness of the frontal and maxillary sinuses.
- ◆ Sinus X-rays reveal cloudiness in the affected sinus, air and fluid, and any thickening of the mucosal lining.
- ◆ Transillumination is a simple diagnostic tool that involves shining a light into the patient's mouth with the lips closed around it. Infected sinuses look dark and normal sinuses transilluminate.
- ◆ Ultrasound, CT scan, magnetic resonance imaging, and X-rays aid in diagnosing suspected complications.

## Treatment

Local decongestants usually are tried before systemic decongestants; steam inhalation may also be helpful. Antibiotics are necessary to combat purulent or persistent infection. Amoxicillin and amoxicillin/clavulanate potassium are usually the antibiotics of choice. Other possible therapy includes cefixime for responsive infections or if beta-lactamase-producing bacteria are present. Because sinusitis is a deep-seated infection, antibiotics should be given for 10 days to 2 weeks. Azithromycin is given for 5 days and may need to be repeated immediately. Local applications of heat may help to relieve pain and congestion. In subacute sinusitis, antibiotics and decongestants may be helpful.

Treatment for allergic sinusitis must include treatment for allergic rhinitis—avoidance measures, administration of antihistamines, identification of allergens by skin testing, and desensitization by immunotherapy. Severe allergic symptoms may require treatment with corticosteroids and epinephrine.

In both chronic sinusitis and hyperplastic sinusitis, using antihistamines, antibiotics, and a steroid nasal spray may relieve pain and congestion. If subacute infection persists, the sinuses may be irrigated. If irrigation fails to relieve symptoms, endoscopic sinus surgery may be required to obtain a histologic diagnosis, remove polyps, and provide adequate ventilation of the infected sinuses. Partial or total resection of the middle turbinate as well as more radical procedures, such as total sphenoethmoidectomy, may be performed.

## Special Considerations

- ◆ Enforce bed rest, and encourage the patient to drink plenty of fluids to promote drainage. Don't elevate the head of the bed by more than 30 degrees.
- ◆ To relieve pain and promote drainage, apply warm compresses continuously, or four times daily for 2-hour intervals. Also, give analgesics and antihistamines as needed.
- ◆ Watch for and report complications, such as vomiting, chills, fever, edema of the forehead or eyelids, blurred or double vision, and personality changes.
- ◆ If surgery is necessary, tell the patient what to expect postoperatively: nasal packing will be in place for 12 to 24 hours following surgery; he'll have to breathe through the mouth and won't be able to blow the nose. After surgery, monitor for excessive drainage or bleeding and watch for complications.
- ◆ To prevent edema and promote drainage, place the patient in semi-Fowler position. To relieve edema and pain and to minimize bleeding, apply ice compresses or a rubber glove filled with ice chips over the nose and iced saline gauze over the eyes. Continue these measures for 24 hours.
- ◆ Frequently change the mustache dressing or drip pad, and record the consistency, amount, and color of drainage (expect scant, bright red, and clotty drainage).

- ◆ Because the patient will be breathing through their mouth, provide meticulous mouth care.
- ◆ Tell the patient that even after the packing is removed, nose blowing may cause bleeding and swelling. If the patient is a smoker, instruct them not to smoke for at least 2 or 3 days after surgery.
- ◆ Tell the patient to finish the prescribed antibiotics, even if their symptoms disappear.

## NASAL POLYPS

Benign and edematous growths, nasal polyps are usually multiple, mobile, and bilateral. Nasal polyps may become large and numerous enough to cause nasal distention and enlargement of the bony framework, possibly occluding the airway.

### Causes and Incidence

Nasal polyps are usually produced by the continuous pressure resulting from a chronic allergy that causes prolonged mucous membrane edema in the nose and sinuses. Other predisposing factors include chronic sinusitis, chronic rhinitis, and recurrent nasal infections.

Nasal polyps are more common in adults than in children and tend to recur. They're also commonly seen in patients with long-term allergic rhinitis and in patients with the aspirin triad (aspirin sensitivity, asthma, and nasal polyps). About 1 in 4 people with cystic fibrosis have nasal polyps.

### Complication

- ◆ Airway obstruction

### Signs and Symptoms

Nasal obstruction is the primary indication of nasal polyps. Such obstruction causes anosmia, a sensation of fullness in the face, nasal discharge, headache, and shortness of breath. Associated clinical features are usually the same as those of allergic rhinitis.

### Diagnosis

Diagnosis of nasal polyps is aided by the following tests.

- ◆ Examination with a nasal speculum shows a dry, red surface, with clear or gray growths. Large growths may resemble tumors.
- ◆ X-rays of sinuses and nasal passages reveal soft-tissue shadows over the affected areas.



**PEDIATRIC TIP** Nasal polyps in children require further testing to rule out cystic fibrosis and Peutz–Jeghers syndrome.

## Treatment

Intranasal glucocorticoids are the treatment of choice. Direct injection into the polyps may temporarily reduce the polyp. A short course of oral corticosteroids (such as prednisone) may be beneficial. Treatment for the underlying cause may include nasal antihistamines to control allergy, and antibiotic therapy if infection is present. Local application of an astringent shrinks hypertrophied tissue.

Surgical treatment should be considered after medical management has failed. A polypectomy is usually performed under a local anesthetic and the use of surgical lasers is becoming more popular; however, patients should be warned that nasal polyps have high recurrence rates. Continued recurrence may require surgical opening of the ethmoid, sphenoid, and maxillary sinuses and evacuation of diseased tissue.

## Special Considerations

- ◆ Administer antihistamines, as ordered, for the patient with allergies. Prepare the patient for scheduled surgery by telling them what to expect postoperatively, such as nasal packing for 1 to 2 days after surgery.

After surgery:

- ◆ Watch for excessive bleeding or other drainage, and promote patient comfort.
- ◆ Elevate the head of the bed to facilitate breathing, reduce swelling, and promote adequate drainage. Change the mustache dressing or drip pad, as needed, and record the consistency, amount, and color of nasal drainage.
- ◆ Intermittently apply ice compresses over the nostrils to lessen swelling, prevent bleeding, and relieve pain.

- ◆ If nasal bleeding occurs—most likely after packing is removed—sit the patient upright, monitor vital signs, and advise not to swallow blood. Compress the outside of the nose against the septum for 10 to 15 minutes. If bleeding persists, nasal packing may be necessary.



**PREVENTION** *Instruct patients with allergies to avoid exposure to allergens and to take antihistamines at the first sign of an allergic reaction. Also, advise them to avoid overuse of nose drops and sprays.*

## NASAL PAPILLOMAS

A papilloma is a benign epithelial tissue overgrowth within the intranasal mucosa. Inverted papillomas grow into the underlying tissue, usually at the junction of the antrum and the maxillary sinus; they generally occur singly but sometimes are associated with squamous cell cancer. Exophytic papillomas, which also tend to occur singly, arise from epithelial tissue, commonly on the surface of the nasal septum.

### Pathophysiology

Inverted papilloma is a benign epithelial growth in the underlying stroma of the nasal cavity and paranasal sinuses. The pathogenesis of this lesion remains unclear, although allergy, chronic sinusitis, and viral infections have been suggested as possible causes.

### Causes and Incidence

A papilloma may arise as a benign precursor of a neoplasm or as a response to tissue injury or viral infection, but its cause is unknown. Both types of papillomas are most prevalent in males. Recurrence is common, even after surgical excision.

### Complications

- ◆ Severe respiratory distress (rare)
- ◆ Nasal drainage
- ◆ Infection

### Signs and Symptoms

Both inverted and exophytic papillomas typically produce symptoms related to unilateral nasal obstruction—congestion, postnasal drip, headache, shortness of breath, dyspnea, and, rarely, severe respiratory distress, nasal drainage, and infection. Epistaxis is most likely to occur with exophytic papillomas. Occasionally hemorrhage may be the presenting symptom.

## Diagnosis

On examination of the nasal mucosa, inverted papillomas usually appear large, bulky, highly vascular, and edematous; color varies from dark red to gray; and consistency, from firm to friable. Exophytic papillomas are usually raised, firm, and rubbery; pink to gray; and securely attached by a broad or pedunculated base to the mucous membrane.



**PEDIATRIC TIP** *Juvenile angiofibroma is a benign vascular tumor that arises in the nasopharynx and occurs most commonly in adolescent males. Nasal obstruction and hemorrhage may occur as with nasal papillomas. Any adolescent male who continues to have recurrent episodes of epistaxis should be assessed for juvenile angiofibroma. Medical management involves surgical excision, with preoperative embolization to reduce bleeding.*



**CONFIRMING DIAGNOSIS** *Tissue biopsy followed by histologic examination of excised tissue confirms the diagnosis.*

## Treatment

The most effective treatment is wide surgical excision or diathermy, with careful inspection of adjacent tissues and sinuses to rule out extension. The use of surgical lasers is becoming more popular. Ibuprofen or acetaminophen and decongestants may relieve symptoms.

## Special Considerations

- ◆ If bleeding occurs, have the patient sit upright, and expectorate blood into an emesis basin. Compress both sides of the nose against the septum for 10 to 15 minutes, and apply ice compresses to the nose. If the bleeding doesn't stop, notify the physician.



**ALERT** Check for airway obstruction. Place your hand under the patient's nostrils to assess air exchange and watch for signs of mild shortness of breath.

- ◆ If surgery is scheduled, tell the patient what to expect postoperatively. Instruct the patient not to blow the nose. (Packing is usually removed 12 to 24 hours after surgery.)
- ◆ Postoperatively, monitor vital signs and respiratory status. Use pulse oximetry to monitor oxygen saturation levels. As needed, administer analgesics and facilitate breathing with a cool-mist vaporizer. Provide mouth care.
- ◆ Frequently change the mustache dressing or drip pad, to ensure proper absorption of drainage. Record the type and amount of drainage. While the nasal packing is in place, expect scant, usually bright red, clotted drainage. Remember that the amount of drainage typically increases for a few hours after the packing is removed.
- ◆ Because papillomas tend to recur, tell the patient to seek medical attention at the first sign of nasal discomfort, discharge, or congestion that doesn't subside with conservative treatment.
- ◆ Encourage regular follow-up visits to detect early signs of recurrence.

## ADENOID HYPERPLASIA

A fairly common childhood condition, adenoid hyperplasia (also known as *adenoid hypertrophy*) is enlargement of the lymphoid tissue of the nasopharynx. Normally, adenoidal tissue is small at birth ( $\frac{3}{4}$ " to  $1\frac{1}{4}$ " [2 to 3 cm]), grows until the child reaches adolescence, and then begins to slowly atrophy. In adenoid hyperplasia, however, this tissue continues to grow. Enlarged adenoids commonly accompany tonsillitis.

### Causes and Incidence

The cause of adenoid hyperplasia is unknown, but contributing factors may include heredity, chronic infection, chronic nasal congestion, persistent allergy, insufficient aeration, and inefficient nasal breathing. Inflammation resulting from repeated infection increases the patient's risk of respiratory obstruction.

### Complications

- ◆ Otitis media
- ◆ Conductive hearing loss
- ◆ Sinusitis
- ◆ Cor pulmonale
- ◆ Pulmonary arterial hypertension

## Signs and Symptoms

Typically, adenoid hyperplasia produces symptoms of respiratory obstruction, especially mouth breathing, snoring at night, and frequent, prolonged nasal congestion. Persistent mouth breathing during the formative years produces voice alteration and distinctive changes in facial features—a slightly elongated face, open mouth, highly arched palate, shortened upper lip, and vacant expression.



**PEDIATRIC TIP** *Occasionally, the child is incapable of mouth breathing, snores loudly at night, and may eventually show effects of nocturnal respiratory insufficiency (sleep apnea), such as intercostal retractions and nasal flaring.*

## Diagnosis



**CONFIRMING DIAGNOSIS** Nasopharyngoscopy or rhinoscopy confirms adenoid hyperplasia by allowing visualization of abnormal tissue. Lateral pharyngeal X-rays show an obliterated nasopharyngeal air column.

## Treatment

Adenoidectomy is the treatment of choice for adenoid hyperplasia and is commonly recommended for the patient with prolonged mouth breathing, nasal speech, adenoid facies, recurrent otitis media, constant nasopharyngitis, and nocturnal respiratory distress. This procedure usually eliminates recurrent nasal infections and ear complications, and reverses any secondary hearing loss.

## Special Considerations

Care requires sympathetic preoperative care and diligent postoperative monitoring.

Before surgery, do the following.

- ◆ Describe the facility routine, and arrange for the patient and their parents to tour relevant areas.
- ◆ Explain adenoidectomy to the child, using illustrations if necessary, and detail the recovery process. Advise them that they'll probably need to be hospitalized. If facility protocol allows, encourage one parent to stay with the child and participate in their care.

After surgery, take these steps.



**ALERT** *Maintain a patent airway. Position the child on their side, with their head down, to prevent aspiration of draining secretions.*

*Frequently check the throat for bleeding. Be alert for vomiting of old, partially digested blood (coffee-ground vomitus). Closely monitor vital signs, and report excessive bleeding, rise in pulse rate, drop in blood pressure, tachypnea, and restlessness.*

- ◆ If no bleeding occurs, offer cracked ice or water when the patient is fully awake.
- ◆ Tell the parents that their child may temporarily have a nasal voice.

## VELOPHARYNGEAL INSUFFICIENCY

Velopharyngeal insufficiency results from failure of the velopharyngeal sphincter to close properly during speech, giving the voice a hypernasal quality and permitting nasal emission (air escape during pronunciation of consonants).

### Causes and Incidence

Velopharyngeal insufficiency can result from an inherited palate abnormality, or it can be acquired from tonsillectomy, adenoidectomy, or palatal paresis. It commonly occurs in people who undergo cleft palate surgery and those with submucous cleft palates. Middle ear disease and hearing loss frequently accompany this disorder.

### Pathophysiology

Velopharyngeal dysfunction (VPD) is a generic term, which describes a set of disorders resulting in the leakage of air into the nasal passages during

speech production. As a result, speech samples can demonstrate hypernasality, nasal emissions, and poor intelligibility. The finding of VPD can be secondary to several causes: anatomic, musculoneuronal, or behavioral/mislearning. To identify the etiology of VPD, patients must undergo a thorough velopharyngeal assessment comprised of perceptual speech evaluation and functional imaging, including video nasendoscopy and speech videofluoroscopy. These studies are then evaluated by a multidisciplinary team of specialists, who can decide on an optimal course for patient management. A treatment plan is developed and may include speech therapy, use of a prosthetic device, and/or surgical intervention. Different surgical options are discussed, including posterior pharyngeal flap, sphincter pharyngoplasty, Furlow palatoplasty, palatal re-repair, and posterior pharyngeal wall augmentation.

## Complication

- ◆ Airway obstruction

## Signs and Symptoms

Generally, this condition causes unintelligible speech, marked by hypernasality, nasal emission, poor consonant definition, and a weak voice. The patient experiences dysphagia and, if velopharyngeal insufficiency is severe, may regurgitate through the nose.

## Diagnosis

Fiberoptic nasopharyngoscopy, which permits monitoring of velopharyngeal patency during speech, suggests this diagnosis. Ultrasound scanning, which shows air-tissue overlap, reflects the degree of velopharyngeal sphincter incompetence (an opening  $>20\text{ mm}^2$  results in unintelligible speech). Videofluoroscopy simultaneously records the movement of the velopharyngeal sphincter and the patient's speech.

## Treatment

Treatment consists of corrective surgery, usually at age 6 or 7. The preferred surgical method is the pharyngeal flap procedure, which diverts a tissue flap from the pharynx to the soft palate. Children with

velopharyngeal insufficiency shouldn't have adenoidectomy except in cases of life-threatening obstruction.

Other appropriate surgical procedures include:

- ◆ augmentation pharyngoplasty, which narrows the velopharyngeal opening by enlarging the pharyngeal wall with a retropharyngeal implant
- ◆ palatal push-back, which separates the hard and soft palates to allow insertion of an obturator, thus lengthening the soft palate
- ◆ pharyngoplasty, which rotates pharyngeal flaps to lengthen the soft palate and narrow the pharynx
- ◆ velopharyngeal sphincter reconstruction, which uses free muscle implantation to reconstruct the sphincter

Surgery eliminates hypernasality and nasal emission, but speech abnormalities persist and usually necessitate speech therapy. Immediate postoperative therapy includes antibiotics and a clear, liquid diet for the first 3 days, followed by a soft diet for 2 weeks.

## **Special Considerations**

- ◆ After surgery for velopharyngeal insufficiency, maintain a patent airway (nasopharynx edema may obstruct the airway). Position the patient on their side, and suction the dependent side of their mouth, avoiding the pharynx.
- ◆ Control postoperative agitation, which may provoke pharyngeal bleeding, with sedation, as ordered.
- ◆ Administer high-humidity oxygen as ordered.
- ◆ Monitor vital signs frequently, and report any changes immediately. Observe for bleeding from the mouth or nose. Check intake and output, and watch for signs of dehydration.
- ◆ Advise the patient that preoperative and postoperative speech therapy require time and effort, but with persistence and practice, speech will improve. Before discharge, emphasize the importance of completing the prescribed antibiotic therapy.

## **Throat**

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### **PHARYNGITIS**

The most common throat disorder, pharyngitis is an acute or chronic inflammation of the pharynx. It frequently accompanies the common cold.

## Causes and Incidence

Pharyngitis is usually caused by a virus. The most common bacterial cause is group A beta-hemolytic streptococci. Other common causes include *Mycoplasma* and *Chlamydia*. In up to 30% of cases, no organism is identified.

Pharyngitis is widespread among adults who live or work in dusty or very dry environments, use their voices excessively, habitually use tobacco or alcohol, or suffer from chronic sinusitis, persistent coughs, or allergies.

## Pathophysiology

Pharyngitis is an inflammatory illness of the mucous membranes and underlying structures of the throat (pharynx). Inflammation usually involves the nasopharynx, uvula, soft palate, and tonsils. The illness can be caused by bacteria, viruses, mycoplasmas, fungi, and parasites and by recognized diseases of uncertain causes. Infection by *Streptococcus* bacteria may be a complication arising from a common cold. The symptoms of streptococcal pharyngitis (commonly known as strep throat) are generally redness and swelling of the throat, a pustulant fluid on the tonsils or discharged from the mouth, extremely sore throat that is felt during swallowing, swelling of lymph nodes, and a slight fever; sometimes in children there are abdominal pain, nausea, headache, and irritability. Diagnosis is established by a detailed medical history and by physical examination; the cause of pharyngeal inflammation can be determined by throat culture. Usually only the symptoms can be treated—with throat lozenges to control sore throat and acetaminophen or aspirin to control fever. If a diagnosis of streptococcal infection is established by culture, appropriate antibiotic therapy, usually with penicillin, is instituted. Within approximately 3 days the fever leaves; the other symptoms may persist for another 2 to 3 days.

## Complications

- ◆ Otitis media
- ◆ Sinusitis
- ◆ Mastoiditis
- ◆ Rheumatic fever

- ◆ Nephritis

## Signs and Symptoms

Pharyngitis produces a sore throat and slight difficulty in swallowing. Swallowing saliva is usually more painful than swallowing food. Pharyngitis may also cause the sensation of a lump in the throat as well as a constant, aggravating urge to swallow. Associated features may include mild fever, headache, muscle and joint pain, coryza, and rhinorrhea. Uncomplicated pharyngitis usually subsides in 3 to 10 days.



**PEDIATRIC TIP** *More than 90% of cases of sore throat and fever in children are of viral origin. Associated symptoms usually include runny nose and nonproductive cough.*

## Diagnosis

Physical examination of the pharynx reveals generalized redness and inflammation of the posterior wall, and red, edematous mucous membranes studded with white or yellow follicles. Exudate is usually confined to the lymphoid areas of the throat, sparing the tonsillar pillars. Bacterial pharyngitis usually produces a large amount of exudate.

A throat culture may be performed to identify bacterial organisms that may be the cause of the inflammation.

## Treatment

Treatment for acute viral pharyngitis is usually symptomatic and consists mainly of rest, warm saline gargles, throat lozenges containing a mild anesthetic, plenty of fluids, and analgesics as needed. If the patient can't swallow fluids, I.V. hydration may be required.

Suspected bacterial pharyngitis requires rigorous treatment with penicillin or another broad-spectrum antibiotic because *Streptococcus* is the chief infecting organism. Antibiotic therapy should continue for 48 hours until culture results are back. If the culture (or a rapid strep test) is positive for group A beta-hemolytic streptococci, or if bacterial infection is suspected despite negative culture results, penicillin therapy should be continued for 10 days. This is to prevent the sequelae of acute rheumatic fever.

Chronic pharyngitis requires the same supportive measures as acute pharyngitis but with greater emphasis on eliminating the underlying cause, such as an allergen. Preventive measures include adequate humidification and avoiding excessive exposure to air-conditioning. In addition, the patient should be urged to stop smoking.

## Special Considerations

- ◆ Administer analgesics and warm saline gargles, as ordered and as appropriate.
- ◆ Encourage the patient to drink plenty of fluids. Scrupulously monitor intake and output, and watch for signs of dehydration.
- ◆ Provide meticulous mouth care to prevent dry lips and oral pyoderma, and maintain a restful environment.
- ◆ Obtain throat cultures, and administer antibiotics as needed. If the patient has acute bacterial pharyngitis, emphasize the importance of completing the full course of antibiotic therapy.
- ◆ Teach the patient with chronic pharyngitis how to minimize sources of throat irritation in the environment, such as by using a bedside humidifier.
- ◆ Refer the patient to a self-help group to stop smoking if appropriate.
- ◆ Children attending school should receive at least 24 hours of therapy before being allowed to return to school.
- ◆ If the patient has exhibited three or more documented bacterial infections within 6 months, consider daily penicillin prophylaxis during the winter months. Also, consider treatment of carriers who live in closed or semiclosed communities.

## TONSILLITIS

Tonsillitis—inflammation of the tonsils—can be acute or chronic. The uncomplicated acute form usually lasts 4 to 6 days. The presence of proven chronic tonsillitis justifies tonsillectomy, the only effective treatment. Tonsils tend to hypertrophy during childhood and atrophy after puberty.

## Causes and Incidence

Tonsillitis generally results from infection with group A beta-hemolytic streptococci but can result from other bacteria or viruses or from oral

anaerobes. It commonly affects children between ages 5 and 10.

## **Pathophysiology**

Tonsillitis is an inflammatory infection of the tonsils caused by invasion of the mucous membrane by microorganisms, usually hemolytic streptococci or viruses. The symptoms are sore throat, difficulty in swallowing, fever, malaise, and enlarged lymph nodes on both sides of the neck. The infection lasts about 5 days. The treatment includes bed rest until the fever has subsided, isolation to protect others from the infection, and warm throat irrigations or gargles with a mild antiseptic solution. Antibiotics or sulfonamides or both are prescribed in severe infections to prevent complications.

## **Complications**

- ◆ Chronic upper airway obstruction
- ◆ Sleep apnea
- ◆ Cor pulmonale
- ◆ Failure to thrive
- ◆ Eating or swallowing disorders
- ◆ Febrile seizures
- ◆ Otitis media
- ◆ Cardiac valvular disease
- ◆ Peritonsillar abscesses
- ◆ Bacterial endocarditis
- ◆ Cervical lymph node abscesses

## **Signs and Symptoms**

Acute tonsillitis commonly begins with a mild to severe sore throat. A very young child, unable to describe a sore throat, may stop eating. Tonsillitis may also produce dysphagia, fever, swelling and tenderness of the lymph glands in the submandibular area, muscle and joint pain, chills, malaise, headache, and pain (frequently referred to the ears). Excess secretions may elicit the complaint of a constant urge to swallow; the back of the throat may feel constricted. Such discomfort usually subsides after 72 hours.

Chronic tonsillitis produces a recurrent sore throat and purulent drainage in the tonsillar crypts. Frequent attacks of acute tonsillitis may also occur.

Complications include obstruction from tonsillar hypertrophy and peritonsillar abscess.

## Diagnosis



**CONFIRMING DIAGNOSIS** *Diagnostic confirmation requires a thorough throat examination that reveals:*

- ◆ *generalized inflammation of the pharyngeal wall*
- ◆ *swollen tonsils that project from between the pillars of the fauces and exude white or yellow follicles*
- ◆ *purulent drainage when pressure is applied to the tonsillar pillars*
- ◆ *possible edematous and inflamed uvula*

Culture may determine the infecting organism and indicate appropriate antibiotic therapy. Leukocytosis is also usually present. Differential diagnosis rules out infectious mononucleosis and diphtheria.

## Treatment

Treatment for acute tonsillitis requires rest, adequate fluid intake, administration of ibuprofen or acetaminophen, and, for bacterial infection, antibiotics. When the causative organism is group A beta-hemolytic streptococcus, penicillin is the drug of choice (another broad-spectrum antibiotic may be substituted). Most oral anaerobes also respond to penicillin. To prevent complications, antibiotic therapy should continue for 10 to 14 days.

Chronic tonsillitis or the development of complications (obstructions from tonsillar hypertrophy, peritonsillar abscess) may require a tonsillectomy, but only after the patient has been free from tonsillar or respiratory tract infections for 3 to 4 weeks.

## Special Considerations

- ◆ Despite dysphagia, urge the patient to drink plenty of fluids, especially if the patient has a fever. Offer a child ice cream and flavored drinks and ices. Suggest gargling with warm salt water to soothe the throat, unless it exacerbates pain. Make sure the patient and parents understand the importance of completing the prescribed course of antibiotic therapy.

- ◆ Before tonsillectomy, explain to the adult patient that a local anesthetic prevents pain but allows a sensation of pressure during surgery. Warn the patient to expect considerable throat discomfort and some bleeding postoperatively. Watch for continuous swallowing, a sign of heavy bleeding.
- ◆ Postoperatively, maintain a patent airway. To prevent aspiration, place the patient on their side. Monitor vital signs frequently, and check for bleeding. Immediately report excessive bleeding, increased pulse rate, or dropping blood pressure. After the patient is fully alert and the gag reflex has returned, allow them to drink water. Later, urge them to drink plenty of nonirritating fluids, to ambulate, and to take frequent deep breaths to prevent pulmonary complications. Give pain medication as needed.
- ◆ Before discharge, provide the patient or their parents with written instructions on home care. Tell them to expect a white scab to form in the throat between 5 and 10 days postoperatively, and to report bleeding, ear discomfort, or a fever that lasts longer than 3 days.



**PEDIATRIC TIP** *For the pediatric patient, keep your explanation simple and nonthreatening. Show the patient the operating and recovery areas, and briefly explain the facility routine. Most facilities allow one parent to stay with the child.*

## THROAT ABSCESES

Throat abscesses may be peritonsillar (quinsy) or retropharyngeal. Peritonsillar abscesses form in the connective tissue space between the tonsil capsule and the constrictor muscle of the pharynx. Retropharyngeal abscesses, or abscesses of the potential space, form between the posterior pharyngeal wall and the prevertebral fascia. With treatment, the prognosis for both types of abscesses is good.

### Causes and Incidence

*Peritonsillar abscess* is a complication of acute tonsillitis, usually after streptococcal or staphylococcal infection. It occurs more commonly in adolescents and young adults than in children.

*Acute retropharyngeal abscess* results from infection in the retropharyngeal lymph glands, which may follow an upper respiratory tract

bacterial infection. Most common pathogens are beta-hemolytic *Streptococcus* and *S. aureus*. These lymph glands begin to atrophy after age 2. Acute retropharyngeal abscess most commonly affects infants and children younger than age 2.

*Chronic retropharyngeal abscess* may result from tuberculosis of the cervical spine (Pott disease) and may occur at any age.

## Pathophysiology

Peritonsillar abscess, the most common deep infection of the head and neck that occurs in adults, is typically formed by a combination of aerobic and anaerobic bacteria. The presenting symptoms include fever, throat pain, and trismus. Ultrasonography and computed tomographic scanning are useful in confirming a diagnosis. Needle aspiration remains the gold standard for diagnosis and treatment of peritonsillar abscess. After performing aspiration, appropriate antibiotic therapy (including penicillin, clindamycin, cephalosporins, or metronidazole) must be initiated. In advanced cases, incision and drainage or immediate tonsillectomy may be required.

## Complications

- ◆ Airway obstruction
- ◆ Cellulitis
- ◆ Endocarditis
- ◆ Pericarditis
- ◆ Pleural effusion
- ◆ Pneumonia

## Signs and Symptoms

Key symptoms of peritonsillar abscess include severe throat pain, occasional ear pain on the same side as the abscess, and tenderness of the submandibular gland. Dysphagia causes drooling. Trismus may occur as a result of the spread of edema and infection from the peritonsillar space to the pterygoid muscles. Other effects include fever, chills, malaise, rancid breath, nausea, muffled speech, dehydration, cervical adenopathy, and localized or systemic sepsis.

Clinical features of retropharyngeal abscess include pain, dysphagia, fever, and, when the abscess is located in the upper pharynx, nasal obstruction; with a low-positioned abscess, dyspnea, progressive inspiratory

stridor (from laryngeal obstruction), neck hyperextension, and, in children, drooling and muffled crying occur. Other symptoms in children may include gurgling respirations, dyspnea and dysphagia, respiratory symptoms, and fever. A very large abscess may press on the larynx, causing edema, or may erode into major vessels, causing sudden death from asphyxia or aspiration.

## Diagnosis

Diagnosis of peritonsillar abscess usually begins with a patient history of bacterial pharyngitis. Examination of the throat shows swelling of the soft palate on the abscessed side, with displacement of the uvula to the opposite side; red, edematous mucous membranes; and tonsil displacement toward the midline. Culture may reveal streptococcal or staphylococcal infection.

Diagnosis of retropharyngeal abscess is based on patient history of nasopharyngitis or pharyngitis and on physical examination revealing a soft, red bulging of the posterior pharyngeal wall. X-rays show the larynx pushed forward and a widened space between the posterior pharyngeal wall and vertebrae. If neck pain or stiffness occurs, look for extension to the epidural space or the cervical vertebrae. Culture and sensitivity tests isolate the causative organism and reveal the appropriate antibiotic.

## Treatment

For early-stage peritonsillar abscess, large doses of penicillin or another broad-spectrum antibiotic is necessary. If the patient is immunocompromised or has been repeatedly hospitalized, antibiotic therapy should include coverage for staphylococci and gram-negative organisms. For late-stage abscess, with cellulitis of the tonsillar space, primary treatment is usually incision and drainage under a local anesthetic, followed by antibiotic therapy for 7 to 10 days. Tonsillectomy, scheduled no sooner than 1 month after healing, prevents recurrence but is recommended only after several episodes.

In acute retropharyngeal abscess, the primary treatment is incision and drainage through the pharyngeal wall. It's considered a surgical emergency. In chronic retropharyngeal abscess, drainage is performed through an external incision behind the sternomastoid muscle. During incision and drainage, strong, continuous mouth suction is necessary to prevent aspiration of pus, and the head should be kept down. Postoperative drug

therapy includes I.V. antibiotics (usually penicillin or clindamycin) and analgesics.

## Special Considerations



**ALERT** *Be alert for signs of respiratory obstruction (inspiratory stridor, dyspnea, retractions and nasal flaring, increasing restlessness, and cyanosis). Keep emergency airway equipment nearby.*

- ◆ Explain the drainage procedure to the patient and their parents. Because the procedure is usually done under local anesthesia, the patient may be apprehensive.
- ◆ Assist with incision and drainage. To allow easy expectoration and suction of pus and blood, place the patient in a semirecumbent or sitting position.

After incision and drainage:

- ◆ Give antibiotics, analgesics, and antipyretics, as ordered. Stress the importance of completing the full course of prescribed antibiotic therapy.
- ◆ Monitor vital signs, and report significant changes or bleeding. Assess pain, and treat accordingly.
- ◆ If the patient is unable to swallow, ensure adequate hydration with I.V. therapy. Monitor fluid intake and output, and watch for dehydration.
- ◆ Provide meticulous mouth care. Apply petroleum jelly to the patient's lips. Promote healing with warm saline gargles or throat irrigations for 24 to 36 hours after incision and drainage. Encourage adequate rest.



**PREVENTION** *Encourage early treatment of tonsillitis.*

## VOCAL CORD PARALYSIS

Vocal cord paralysis results from disease of, or injury to, the superior or, most commonly, the recurrent laryngeal nerve. It may also be congenital.

## Causes and Incidence

Vocal cord paralysis commonly results from the accidental severing of the recurrent laryngeal nerve, or of one of its extralaryngeal branches, during thyroidectomy. Other causes include pressure from a thoracic aortic

aneurysm or from an enlarged atrium (in patients with mitral stenosis), bronchial or esophageal carcinoma, hypertrophy of the thyroid gland, trauma (such as neck injuries) and intubation, and neuritis due to infections or metallic poisoning. Vocal cord paralysis can also result from hysteria and, rarely, lesions of the central nervous system.

## Pathophysiology

Unilateral vocal fold paralysis occurs from a dysfunction of the recurrent laryngeal or vagus nerve innervating the larynx. It causes a characteristic breathy voice often accompanied by swallowing disability, a weak cough, and the sensation of shortness of breath. This is a common cause of neurogenic hoarseness.

## Complications

- ◆ Airway obstruction
- ◆ Respiratory failure

## Signs and Symptoms

Unilateral paralysis, the most common form, may cause vocal weakness and hoarseness. Bilateral paralysis typically produces vocal weakness and incapacitating airway obstruction if the cords become paralyzed in the adducted position.



**PEDIATRIC TIP** Children may present with hoarseness, aspiration, and stridor. If the paralysis is unilateral, it typically involves the left recurrent laryngeal nerve. In unilateral paralysis, airway intervention involving intubation and tracheostomy is rarely indicated; it's usually required if the paralysis is bilateral.

## Diagnosis

The patient history and characteristic features suggest vocal cord paralysis.



**CONFIRMING DIAGNOSIS** Visualization by indirect laryngoscopy shows one or both cords fixed in an adducted or partially abducted position and confirms the diagnosis.

X-ray or CT scan detect abnormalities in the mediastinum that may be responsible for the injury.

## Treatment

Treatment for unilateral vocal cord paralysis consists of injection of Teflon into the paralyzed cord, under direct laryngoscopy. This procedure enlarges the cord and brings it closer to the other cord, which usually strengthens the voice and protects the airway from aspiration. Thyroplasty also serves to reposition the vocal cord, but in this procedure an implant is placed through a neck incision. The ansa cervicalis nerve transfer allows for reinnervation of the muscles of the vocal cord. Bilateral cord paralysis in an adducted position necessitates a tracheostomy.

Alternative treatments for adults include endoscopic arytenoidectomy to open the glottis, and lateral fixation of the arytenoid cartilage through an external neck incision. Excision or fixation of the arytenoid cartilage improves airway patency but produces residual voice impairment.

Treatment for hysterical aphonia may include psychotherapy and hypnosis.

## Special Considerations

If the patient chooses direct laryngoscopy and Teflon injection, explain these procedures thoroughly. Tell the patient these measures will improve their voice but won't restore it to normal. Patients are sometimes placed on voice rest for 24 to 48 hours to reduce stress on the vocal cords, which would increase the edema and might lead to airway obstruction.

Many patients with bilateral cord paralysis prefer to keep a tracheostomy instead of having an arytenoidectomy; voice quality is generally better with a tracheostomy alone than after corrective surgery.

If the patient is scheduled to undergo a tracheostomy:

- ◆ Explain the procedure thoroughly, and offer reassurance. Because the procedure is performed under a local anesthetic, the patient may be apprehensive.
- ◆ Teach the patient how to suction, clean, and change the tracheostomy tube.
- ◆ Reassure the patient that they can still speak by covering the lumen of the tracheostomy tube with a finger or a tracheostomy plug.

If the patient elects to have an arytenoidectomy, explain the procedure thoroughly. Advise the patient that the tracheostomy will remain in place until the edema has subsided and the airway is patent.

## VOCAL CORD NODULES AND POLYPS

Vocal cord nodules result from hypertrophy of fibrous tissue and form at the point where the cords come together forcibly. Vocal cord polyps are chronic, subepithelial, edematous masses. Both nodules and polyps have a good prognosis unless continued voice abuse causes recurrence, with subsequent scarring and permanent hoarseness.

### Causes and Incidence

Vocal cord nodules and polyps usually result from voice abuse, especially in the presence of infection. Consequently, they're most common in teachers, singers, and sports fans, and in energetic children (ages 8 to 12) who continually shout while playing. Polyps are common in adults who smoke, live in dry climates, or have allergies.



**PEDIATRIC TIP** *In children, papillomas of the larynx (benign warty growths) are the most common laryngeal neoplasm. Suspected causes include human papillomavirus types 6, 11, and 16. The virus may be acquired during birth because many mothers have a history of condylomata acuminata at the time of delivery.*

### Complication

- ◆ Permanent hoarseness

### Signs and Symptoms

Nodules and polyps inhibit the approximation of vocal cords and produce painless hoarseness. The voice may also develop a breathy or husky quality.

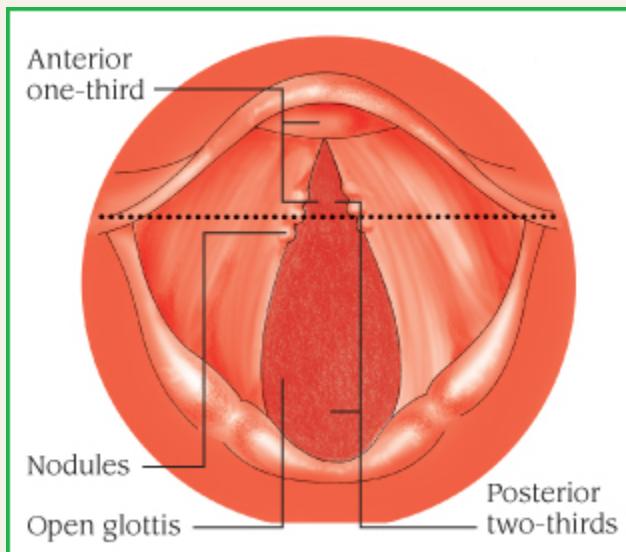
### Diagnosis

Persistent hoarseness suggests vocal cord nodules and polyps; visualization by indirect laryngoscopy confirms it. In the patient with vocal cord nodules, laryngoscopy initially shows small red nodes and, later, white solid nodes

on one or both cords. (See *Vocal cord nodules*.) In the patient with polyps, laryngoscopy reveals unilateral or, occasionally, bilateral, sessile or pedunculated polyps of varying size, anywhere on the vocal cords.

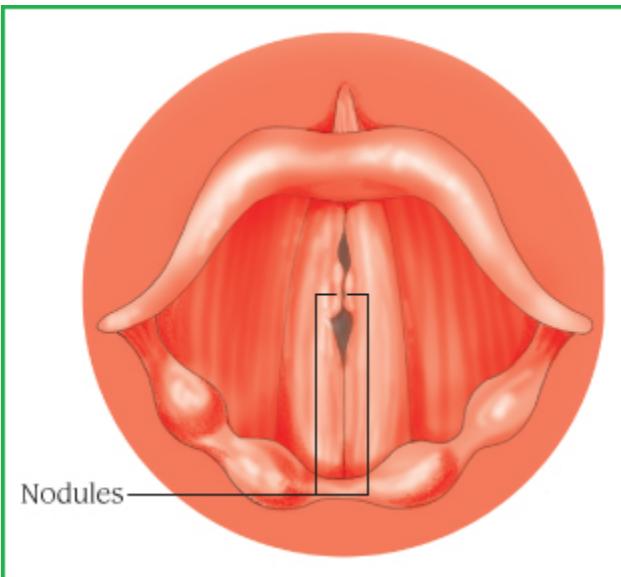
## Vocal Cord Nodules

The most common site of vocal cord nodules is the point of maximal vibration and impact (junction of the anterior one third and the posterior two thirds of the vocal cord).



Vocal Cords Open

Vocal cord nodules affect the voice by inhibiting proper closure of the vocal cords during phonation.



Vocal Cords Closed

## Treatment

Conservative management of small vocal cord nodules and polyps includes humidification, speech therapy (voice rest, training to reduce the intensity and duration of voice production), and treatment for any underlying allergies.

When conservative treatment fails to relieve hoarseness, nodules or polyps require removal under direct laryngoscopy. Microlaryngoscopy may be done for small lesions, to avoid injuring the vocal cord surface. If nodules or polyps are bilateral, excision may be performed in two stages: one cord is allowed to heal before excision of polyps on the other cord. Two-stage excision prevents laryngeal web, which occurs when epithelial tissue is removed from adjacent cord surfaces, and these surfaces grow together.



**PEDIATRIC TIP** *For children, treatment consists of speech therapy. If possible, surgery should be delayed until the child is old enough to benefit from voice training, or until the patient can understand the need to abstain from voice abuse.*

## Special Considerations

- ◆ Postoperatively, stress the importance of resting the voice for 10 to 14 days while the vocal cords heal. Provide an alternative means of communication—Magic Slate, pad and pencil, or alphabet board. Place a sign over the bed to remind visitors that the patient shouldn't talk. Mark the intercom so other facility personnel are aware that the patient can't answer. Minimize the need to speak by trying to anticipate the patient's needs.
- ◆ If the patient is a smoker, encourage them to stop smoking entirely or, at the very least, to refrain from smoking during recovery from surgery.
- ◆ Use a vaporizer to increase humidity and decrease throat irritation.
- ◆ Make sure the patient receives speech therapy after healing if necessary, because continued voice abuse causes recurrence of growths.

## LARYNGITIS

A common disorder, laryngitis is an acute or chronic inflammation of the vocal cords. Acute laryngitis may occur as an isolated infection or as part of a generalized bacterial or viral URTI. Repeated attacks of acute laryngitis produce inflammatory changes associated with chronic laryngitis.



**ALERT** *Several forms of laryngitis occur in children and can lead to significant or fatal respiratory obstruction, such as croup and epiglottitis.*

### Causes and Incidence

Acute laryngitis usually results from infection (primarily viral) or excessive use of the voice, an occupational hazard in certain vocations (e.g., teaching, public speaking, or singing). It may also result from leisure activities (such as cheering at a sports event), inhalation of smoke or fumes, or aspiration of caustic chemicals. Chronic laryngitis may be caused by chronic upper respiratory tract disorders (sinusitis, bronchitis, nasal polyps, or allergy), mouth breathing, smoking, constant exposure to dust or other irritants, and alcohol abuse.

### Pathophysiology

Acute laryngitis is an inflammation of the vocal fold mucosa and larynx that lasts less than 3 weeks. When the etiology of acute laryngitis is infectious, white blood cells remove microorganisms during the healing

process. The vocal folds then become more edematous, and vibration is adversely affected.

## Complications

- ◆ Permanent hoarseness
- ◆ Airway obstruction in severe laryngitis

## Signs and Symptoms

Acute laryngitis typically begins with hoarseness, ranging from mild to complete loss of voice. Associated clinical features include pain (especially when swallowing or speaking), a persistent dry cough, fever, laryngeal edema, and malaise. In chronic laryngitis, persistent hoarseness is usually the only symptom.

## Diagnosis

**Dx CONFIRMING DIAGNOSIS** *Indirect laryngoscopy confirms the diagnosis by revealing red, inflamed, and, occasionally, hemorrhagic vocal cords, with rounded rather than sharp edges and exudate. Bilateral swelling may be present.*

In severe cases or if toxicity is a concern, a culture of the exudate is obtained. Consider 24-hour pH probe testing in chronic laryngitis and GERD. Also consider biopsy in chronic laryngitis in an adult with a history of smoking or alcohol abuse.

## Treatment

Primary treatment consists of resting the voice. For viral infection, symptomatic care includes analgesics and throat lozenges for pain relief. Bacterial infection requires antibiotic therapy. Severe, acute laryngitis may necessitate hospitalization. When laryngeal edema results in airway obstruction, a tracheostomy may be necessary. In chronic laryngitis, effective treatment must eliminate the underlying cause. Antacids or histamine-2 blockers may be used if GERD is the cause. Steam inhalation may also prove beneficial as are smoking cessation, reducing alcohol intake, and job change or modification if warranted.

## **Special Considerations**

- ◆ Explain to the patient why they shouldn't talk, and place a sign over the bed to remind others of this restriction. Provide a Magic Slate or a pad and pencil for communication. Mark the intercom panel so other facility personnel are aware that the patient can't answer. Minimize the need to talk by trying to anticipate the patient's needs.
- ◆ For the patient with a bacterial infection, stress the importance of completing the full course of antibiotic therapy.
- ◆ Suggest that the patient maintain adequate humidification by using a vaporizer or humidifier during the winter, by avoiding air-conditioning during the summer (because it dehumidifies), by using medicated throat lozenges, and by not smoking.
- ◆ Obtain a detailed patient history to help determine the cause of chronic laryngitis. Encourage the patient to modify predisposing habits, especially to stop smoking.
- ◆ Provide the patient with assistance for smoking cessation as well as for modification of other predisposing habits or occupational hazards.

## **JUVENILE ANGIOFIBROMA**

An uncommon disorder, juvenile angiofibroma is a highly vascular, nasopharyngeal tumor made up of masses of fibrous tissue that contain many thin-walled blood vessels. The prognosis is good with treatment.

### **Causes and Incidence**

A type of hemangioma, this tumor grows on one side of the posterior nares and may completely fill the nasopharynx, nose, paranasal sinuses, and, possibly, the orbit. More commonly sessile than polypoid, juvenile angiofibroma is nonencapsulated; it invades surrounding tissue.

Juvenile angiofibroma is typically found in adolescent males and is extremely rare in females. It's associated with nasal obstruction and epistaxis.

### **Pathophysiology**

JNA is a rare benign tumor arising predominantly in the nasopharynx of adolescent males. It is an aggressive neoplasm and shows a propensity for destructive local spread often extending to the base of the skull and into the

cranium. Clinically, however, it is obscure with painless, progressive unilateral nasal obstruction being the common presenting symptom with or without epistaxis and rhinorrhea. Diagnosis of JNA is made by complete history, clinical examination, radiography, nasal endoscopy and by using specialized imaging techniques such as arteriography, CT, and magnetic resonance imaging. Early diagnosis, accurate staging, and adequate treatment are essential in the management of this lesion.

## Complication

- ◆ Secondary anemia

## Signs and Symptoms

Juvenile angiofibroma produces unilateral or bilateral nasal obstruction and severe recurrent epistaxis, usually between ages 7 and 21. Recurrent epistaxis eventually causes secondary anemia. Associated effects include purulent rhinorrhea, facial deformity, and nasal speech. Serous otitis media and hearing loss may result from eustachian tube obstruction.

## Diagnosis

A nasopharyngeal mirror or nasal speculum permits visualization of the tumor. X-rays show a bowing of the posterior wall of the maxillary sinus. Three-plane magnetic resonance imaging and CT scans determine the extent of the tumors, which are seldom limited to the nasopharynx. Angiography determines the size and location of the tumor and shows the source of vascularization.



**ALERT** Tumor biopsy is contraindicated because of the risk of hemorrhage.

## Treatment

Surgical procedures range from avulsion to cryosurgical techniques. Surgical excision is preferred after embolization with Teflon or an absorbable gelatin sponge to decrease vascularization. Whichever surgical method is used, this tumor must be removed in its entirety and not in pieces.

Preoperative hormonal therapy may decrease the tumor's size and vascularity. Blood transfusions may be necessary during avulsion. Radiation therapy produces only a temporary regression in an angiofibroma

but is the treatment of choice if the tumor has expanded into the cranium or orbit. Because the tumor is multilobular and locally invasive, it recurs in about 30% of patients during the first year after treatment, but rarely after 2 years.

## **Special Considerations**

- ◆ Explain all diagnostic and surgical procedures. Provide emotional support; severe epistaxis frightens many people to the point of panic. Monitor hemoglobin levels and HCT for anemia.
- ◆ After surgery, immediately report excessive bleeding. Make sure an adequate supply of typed and crossmatched blood is available for transfusion.
- ◆ Monitor for any change in vital signs. Provide good oral hygiene, and use a bedside vaporizer to raise humidity.
- ◆ During blood transfusion, watch for transfusion reactions, such as fever, pruritus, chills, or a rash. If any of these reactions occur, discontinue the blood transfusion and notify the physician immediately.
- ◆ Teach the patient's family how to apply pressure over the affected area, and instruct them to seek immediate medical attention if bleeding occurs after discharge. Stress the importance of providing adequate humidification at home to keep the nasal mucosa moist.

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