

platelet aggregation and thereby reducing the combined risk of ischemic stroke, MI, or death from vascular disease and may be useful in preventing coronary stent thrombosis.

Revascularization procedures are recommended for persons who do not become ischemia free on medical therapy, especially clients with progressive unstable angina. Surgical intervention, such as PTCA, has been shown to relieve angina, but it does not halt the progress of atherosclerosis. CABG can diminish the probability that ischemia will lead to necrosis and lethal infarction.

For people who are not good candidates for any of the proven procedures or whose angina persists despite angioplasty or bypass surgery, transmyocardial revascularization (TMR) may be recommended. In TMR, a computer-controlled laser drills tiny channels through the wall of the left ventricle while the chamber is filled with oxygenated blood. In theory, this allows blood to flow through the channels to the oxygen-deprived tissue, relieving angina. The openings on the heart's surface scar over quickly, but it is not known how long the channels stay open on the inside of the heart; long-term results remain unknown.

TMR is still considered experimental and may not be available in all centers, but studies consistently report success in relieving severe angina that has been refractory to medical and surgical intervention.¹⁶⁹ This procedure is performed through a 4-inch incision between the ribs and does not require a bypass machine; efforts to develop noninvasive techniques for TMR using fiberoptic catheters are under investigation.

PROGNOSIS. Myocardial ischemia leaves the heart vulnerable to arrhythmias and MI, which can be fatal. About one third of all people who experience angina pectoris die suddenly from MI or arrhythmias. Prognosis depends primarily on left ventricular function (i.e., ejection fraction) but is influenced by type of angina, ability to prevent angina, and severity of underlying disease, such as hypertension or atherosclerosis.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-3

Angina Pectoris

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

See also Special Implications for the Therapist: Atherosclerosis.

Identifying Angina

Referred pain from the external oblique abdominal muscle and the pectoral major muscle can cause the sensation referred to as heartburn in the anterior chest wall, which mimics angina. When active trigger points are present in the left pectoralis major muscle, the referred pain is easily confused with that from coronary insufficiency. Physical therapy to eliminate the trigger points can aid in the diagnostic process.

Anterior chest wall syndrome with localized tenderness of intercostal muscles, Tietze's syndrome with inflammation of the chondrocostal junctions, intercostal neuritis, and cervical or thoracic spine disease involving the dorsal nerve roots can all produce chest pain that mimics angina. Evaluation of range of motion, palpation of soft tissue structures, and analysis of relieving or aggravating factors usually differentiate these conditions from true angina.¹²⁹ Likewise, heartburn from indigestion, hiatal hernia, peptic ulcer, esophageal spasm, and gallbladder disease can also cause angina-like symptoms that require a medical evaluation for an accurate medical diagnosis.

The development of unstable angina also requires immediate medical referral and may be reported as the onset of angina at rest, occurrence of typical angina at a significantly lower level of activity than usual, changes in the typical anginal pattern (e.g., symptoms occurring more frequently), or changes in blood pressure (decrease) or heart rate (increase) with levels of activity previously well tolerated. Educating the public about reducing delays and getting to an emergency department at the earliest signs of heart attack is essential. Reperfusion therapy within the first 60 to 70 minutes of a heart attack can make a significant difference in outcome.

Nitroglycerin

A person experiencing angina should reduce the pace of or, if necessary, stop all activity and sit down for a few minutes until the symptoms disappear. Exercise can be reinitiated at a reduced intensity, and interval-type training may be required (i.e., slow activity alternating with activity requiring more effort). Some experts suggest waiting several hours before resuming exercise. Anyone experiencing angina regularly with exercise or at a lower exertion than in the past may need a medical evaluation.

Nitroglycerin may be used prophylactically 5 minutes before activities likely to precipitate angina. This is especially true in the intervention or exercise setting for the person with chronic, stable, exertional angina. The use of nitroglycerin must be by physician order and cannot be decided solely by the therapist and client.

Clients must be reminded that they are not to alter their prescribed drug schedule without consulting their health care provider and that nitrates should be taken as prescribed. For example, taking sublingual nitroglycerin orally markedly decreases its effectiveness. Clients should be seated when taking nitroglycerin to avoid syncope and falls. For anginal pain or discomfort that is not relieved by rest or relieved by up to three nitroglycerin doses in 10 to 15 minutes (i.e., the initial dose followed by a second dose 5 minutes later and a third dose 5 minutes after the second dose), the physician should be contacted. Until the angina is controlled and coronary blood flow reestablished, the client is at risk of myocardial damage from myocardial ischemia.

Continued.

Nitroglycerin tablets are inactivated by light, heat, air, and moisture, and they should be stored in the refrigerator in an amber container with a tight-fitting cover. Nitroglycerin has a short shelf life and needs to be replaced about every 3 months. A potent nitroglycerin tablet should produce a burning sensation under the tongue when taken sublingually (if it does not, check the expiration date).

Orthostatic Hypotension

Orthostatic hypotension (see discussion later in this chapter) is one of the most common side effects of prophylactic medications for angina. Caution on the part of the therapist is required when exercising or ambulating clients who take these medications. If the person becomes hypotensive, have him or her assume a supine position with legs elevated to increase venous return and to ensure cerebral blood flow.

Extra caution must be taken when placing anyone with orthostatic hypotension and CHF supine with legs elevated because this may overload an already stressed ventricle. Keeping the head elevated and monitoring carefully are required in this circumstance. Support hose may be recommended, and the person should be reminded to change positions slowly to minimize the effects of orthostatic hypotension. Headache, weakness, increasing pulse, or other unusual signs or symptoms should be reported to the physician. In a home health setting, the home should be evaluated for potentially hazardous conditions. All clients should be encouraged to avoid hazardous activities until their condition is stabilized by medication, especially in the presence of dizziness.

Monitoring Vital Signs

Exercise testing should be performed before a client begins an exercise program, but if this has not been accomplished and baseline measurements are unavailable for use in planning exercise, monitor the heart rate and blood pressure and note any accompanying symptoms during exercise (see Appendix B). Exercise and activity should be performed below the anginal threshold. The therapist must document heart rate and blood pressure when the ischemia began (as evidenced by symptoms of angina) to establish these parameters. Angina occurring after an MI is not considered normal and should be reported to the physician. Exercise testing is recommended before a client resumes an exercise program.

Hypertensive Cardiovascular Disease

Hypertensive cardiovascular disease includes hypertensive vascular disease and hypertensive heart disease. Other conditions affecting the heart caused by an underlying pulmonary pathologic condition (e.g., pulmonary hypertension, pulmonary heart disease) are discussed in Chapter 15.

Hypertension (Hypertensive Vascular Disease)

Definition and Overview. Blood pressure is the force exerted against the walls of the arteries and arterioles; diastolic pressure (bottom number) is the pressure in these vessels when the heart is relaxed between beats, and systolic pressure (top number) is the pressure exerted in the arteries when the heart contracts. Between ages 55 and 60 years, diastolic blood pressure often begins to plateau and may even decline, whereas systolic blood pressure often starts to rise.

Hypertension, or high blood pressure, is defined by the World Health Organization (WHO) as a persistent elevation of diastolic blood pressure (higher than 90 mm Hg), systolic blood pressure (higher than 140 mm Hg), or both measured on at least two separate occasions at least 2 weeks apart (i.e., sustained elevation of blood pressure). Recent evidence supports a systolic blood pressure threshold of 140 mm Hg for even "low-risk" individuals. In high-risk persons, there is evidence for lower thresholds.³⁵⁸

Based on epidemiologic data from the Framingham Heart Study, the development of hypertension is neither inevitable nor beneficial; both systolic pressure and diastolic pressure are important determinants of cardiovascular sequelae.

Hypertension can be classified according to type (systolic or diastolic), cause, and degree of severity. Hypertension can also be classified based on risk according to the most recent guidelines (Table 12-8).⁷⁵

Primary (or essential) *hypertension* is also known as idiopathic hypertension and accounts for 90% to 95% of all cases of hypertension. *Secondary hypertension* accounts for only 5% to 10% of cases and results from an identifiable cause. Intermittent elevation of blood pressure interspersed with normal readings is called *labile hypertension* or borderline hypertension. *Malignant hypertension* is a syndrome of markedly elevated blood pressure (diastolic blood pressure of more than 125 mm Hg) with target organ damage (e.g., retinal hemorrhages, papilledema, heart failure, encephalopathy, renal insufficiency). The elevation of systolic blood pressure independently of change in the diastolic blood pressure is now recognized as a medical condition referred to as *isolated systolic hypertension*.

Incidence. The incidence of hypertension varies considerably among different groups in the American population, but it is estimated that one in four adult Americans (50 million) have high blood pressure. Hypertension is twice as prevalent and more severe among blacks than whites. This phenomenon has been attributed to heredity, greater environmental stress, and greater salt intake or salt sensitivity (i.e., responsiveness to changes in sodium balance and extracellular fluid and volume status), although the actual cause is not clear; reduced access to health care increases the prevalence of untreated hypertension.

Blood pressure control rates vary in minority populations and are lowest in Mexican Americans and Native Americans.⁷⁵ Socioeconomic factors and lifestyle may be important barriers to blood pressure control in some minority individuals.

Table 12-8 Classification of Blood Pressure for Adults

	Systolic Blood Pressure	Diastolic Blood Pressure
Normal	<120 mm Hg	<80 mm Hg
Prehypertension	120-139	80-89
Stage 1 hypertension	140-159	90-99
Stage 2 hypertension	≥160	≥100

From The Seventh Report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure. NIH publication no. 03-5233, Bethesda, MD, May 2003, National Heart, Lung, and Blood Institute. Available on-line at www.nhlbi.nih.gov. Accessed April 21, 2008.

The relationship between blood pressure (BP) and risk of cardiovascular disease (CVD) events is continuous, consistent, and independent of other risk factors. The higher the BP, the greater the chance of heart attack, heart failure, stroke, and kidney disease.

For individuals 40-70 yr of age, each 20 mm Hg incremental increase in systolic BP or 10 mm Hg in diastolic BP doubles the risk of CVD across the entire BP range from 115/75 to 185/115 mm Hg.

Classification of Blood Pressure for Children and Adolescents

Normal	<90th percentile; 50th percentile is the midpoint of the normal range
Prehypertension	90th-95th percentile or if blood pressure is greater than 120/80 (even if this figure is <90th percentile)
Stage 1 hypertension	95th-99th percentile + 5 mm Hg
Stage 2 hypertension	>99th percentile + 5 mm Hg

From National Heart, Lung, and Blood Institute: Fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents, *Pediatrics* 114(2):555-576, August 2004.

Hypertension control rates are no longer improving and fell short of the U.S. *Healthy People 2000* goal of blood pressure control in 50% of all hypertensive adults. These goals have now been modified for *Healthy People 2010* to include the following: (1) reduce the proportion of adults ages 20 years and older with high blood pressure from the current incidence (28%) to 16% and (2) increase the proportion of adults ages 18 years and older with high blood pressure whose blood pressure is under control from current rates of 18% to a target rate of 50%.¹⁴⁶

Etiologic and Risk Factors. Primary (essential) hypertension has no established etiology but is probably related to genetics and other risk factors, such as smoking, obesity, high cholesterol levels, and being of black descent. In the recent past, hypertension research shifted strongly in the direction of molecular genetics.

A familial association with hypertension has been documented, possibly attributable to common genetic background, shared environment, or lifestyle habits. Using the candidate gene approach, allelic variants in the genes for angiotensinogen, β_2 -adrenergic receptor, mutation in the β subunit of the epithelial sodium channel, and several others have been identified, but the mecha-

Box 12-7

CAUSES OF SECONDARY HYPERTENSION

- Coarctation of the aorta
- Pheochromocytoma (rare catecholamine-secreting tumor)
- Alcohol abuse
- Pregnancy
- Thyrotoxicosis
- Increased intracranial pressure from tumors or trauma
- Collagen disease
- Endocrine disease
 - Acromegaly
 - Cushing's disease
 - Diabetes mellitus
 - Hypothyroidism
 - Hyperthyroidism
- Renal disease (e.g., connective tissue diseases, diabetic nephropathy)
- Effects of drugs (e.g., oral contraceptives, corticosteroids, cyclosporine, cocaine)
- Acute stress
 - Surgery
 - Psychogenic hyperventilation
 - Alcohol withdrawal
 - Burns
 - Pancreatitis
 - Sickle cell crisis
- Neurologic disorders
 - Brain tumor
 - Respiratory acidosis
 - Encephalitis
 - Sleep apnea
 - Guillain-Barré syndrome
 - Quadriplegia
 - Lead poisoning

Modified from Kaplan NM: Systemic hypertension: mechanisms and diagnosis. In Braunwald E, ed: *Heart disease: a textbook of cardiovascular medicine*, ed 4, Philadelphia, 1992, Saunders.

nisms by which these contribute to hypertension have not been identified.²¹⁰

Small arteries branching from the aorta, called arterioles, regulate blood pressure. Any condition that can narrow the opening of these arterioles can increase the blood pressure in the arteries. A variety of specific diseases or problems, such as chronic renal failure, renal artery stenosis, or endocrine disease, can cause *secondary hypertension* (Box 12-7). The risk for cardiovascular disease in adults with hypertension is determined not only by the level of blood pressure but also by the presence or absence of target organ damage or factors such as smoking, dyslipidemia, and diabetes. *Isolated systolic hypertension* has very distinct causes, often not directly vascular and often related to a specific organ or tissue, such as aortic diseases, heart malformations, or thyrotoxic crisis.

Risk factors for hypertension may be modifiable or nonmodifiable (Box 12-8). The risk of hypertension increases with age as arteries lose elasticity and become less able to relax. Hypertension occurs slightly more often in men than in women and at an earlier age, but after age 50 years, hypertension begins to develop in more women than men. In all groups the incidence of hypertension

Box 12-8**RISK FACTORS OF PRIMARY (ESSENTIAL) HYPERTENSION****Modifiable**

- High sodium intake (causes water retention, increasing blood volume)
- Obesity (associated with increased intravascular volume)
- Diabetes mellitus
- Hypercholesterolemia and increased serum triglyceride levels
- Smoking (nicotine restricts blood vessels)
- Long-term abuse of alcohol (increases plasma catecholamines)
- Continuous emotional stress (stimulates sympathetic nervous system)
- Personality traits (hostility, sense of hopelessness)
- Sedentary lifestyle
- White coat hypertension (see explanation in text)
- Hormonal status (menopause, especially before age 40 years and without hormone replacement therapy; hysterectomy/oophorectomy)

Nonmodifiable

- Family history of cardiovascular disease
- Age (>55 years)
- Gender (male <55 years; female >55 years)
- Ethnicity (black, * Hispanic)

*From a pathogenetic point of view, recent research findings have suggested that β -adrenergic receptor down-regulation is characteristic of hypertension in whites, whereas heightened vascular α -receptor sensitivity or early vascular hypertrophy may be a feature of hypertension in African Americans.³⁰⁷ African Americans demonstrate somewhat reduced blood pressure responses to monotherapy with β -blockers, angiotensin-converting enzyme inhibitors, or angiotensin receptor blockers compared with diuretics or calcium channel blockers. These differential responses are largely eliminated by drug combinations.^{24,75}

increases with age, with a poorer prognosis for people whose hypertension begins at a young age.

White coat hypertension increases the risk of heart disease, because rise in blood pressure occurs in other anxiety-provoking situations as well. Personality traits such as hopelessness and hostility are important factors in cardiovascular disease, including hypertension; research is under way to identify the neuroendocrine and CNS mechanisms underlying these associations and to identify other possible risk-associated personality traits.¹⁰³ Hypertension itself represents a significant risk factor for the development of CAD, stroke, CHF, and renal failure, preceding heart failure in 90% of all cases and increasing in all other associated conditions.

The influence of nonsteroidal antiinflammatory drugs (NSAIDs) on blood pressure in normotensive and hypertensive persons remains in question. At the very least it has been determined that there is always the potential for NSAIDs to interact with antihypertensive agents, most notably diuretics, β -receptor antagonists, and ACE inhibitors.

Given the high prevalence of NSAID use by older adults, especially for conditions such as arthritis, gout, and similar problems, the association between this drug use and blood pressure must be observed carefully (see

Special Implications for the Therapist: Hypertension, section on Medications). Alcohol has been estimated to be responsible for as many as 10% of all cases of hypertension and may be the actual unknown cause of "essential" hypertension.¹¹⁵

Blood pressure is linked to salt intake and modulated by the "salt gene" (angiotensinogen) in some people. Those who are salt sensitive (including those who do not yet have high blood pressure) may have an increased risk of death. Salt sensitivity is a measure of how blood pressure responds to sodium and is independent of other risk factors (including elevated blood pressure) for death from cardiovascular disease.

Inadequate sleep has been identified as a risk factor for hypertension among adults in their fourth to sixth decades who sleep less than 5 hours each night. Short sleep duration is also a risk factor for obesity and diabetes, two conditions commonly linked with hypertension. Men are more likely than women to report getting fewer than 6 hours of sleep, although women are more likely to have trouble falling asleep or getting back to sleep after waking up early.^{102,119}

Pathogenesis. Blood pressure is regulated by two factors: blood flow and peripheral vascular resistance. Blood flow is determined by cardiac output (strength, rate, rhythm of heartbeat; blood volume). The resistance to flow is primarily determined by the diameter of blood vessels and, to a lesser degree, by the viscosity of blood.

Increased peripheral resistance as a result of the narrowing of the arterioles is the single most common characteristic of hypertension. Constriction of the peripheral arterioles may be controlled by two mechanisms, each with several components: (1) sympathetic nervous system activity (autonomic regulation) and (2) activation of the renin-angiotensin system.

In the case of the sympathetic nervous system, norepinephrine is released in response to psychogenic stress or baroreceptor activity. The blood vessels constrict, which increases peripheral resistance. At the same time, epinephrine is secreted by the adrenal medulla, resulting in increased force of cardiac contraction, increased cardiac output, and vasoconstriction.

With prolonged hypertension, the elastic tissue in the arterioles is replaced by fibrous collagen tissue. The thickened arteriole wall becomes less distensible, offering even greater resistance to the flow of blood. This process leads to decreased tissue perfusion, especially in the target organs of high blood pressure (i.e., heart, kidneys, brain). Atherosclerosis is also accelerated in persons with high blood pressure.

Within the renin-angiotensin system, vasoconstriction results in decreased blood flow to the kidney. Whenever blood flow to the kidney diminishes, renin is secreted and angiotensin is formed, causing vasoconstriction within the renal system and increased total peripheral resistance. Angiotensin also stimulates the secretion of aldosterone, which promotes sodium and water retention by the kidney tubules, causing an increase in intravascular volume. All these factors increase blood pressure.

Evidence from animal, clinical, and epidemiologic studies points to an association between high blood pres-

sure and abnormal calcium metabolism, leading to increased calcium loss, secondary activation of the parathyroid gland, increased movement of calcium from bone, and increased risk of urinary tract stones and osteoporosis.⁶²

Clinical Manifestations. Hypertension is frequently asymptomatic; this creates a significant health care risk for affected people. When symptoms do occur, they may include headache (usually occipital and present in the morning, worse on waking, and slowly improving with activity), vertigo, flushed face, spontaneous epistaxis, blurred vision, and nocturnal urinary frequency. Elevated blood pressure when measured, especially in the early stages, may be the only sign of hypertension.

Sleep-disordered breathing is also associated with systemic hypertension in middle-aged and older individuals of both genders and different ethnic backgrounds.²⁴² Progressive hypertension may be characterized by cardiovascular symptoms (dyspnea, orthopnea, chest pain, and leg edema) or cerebral symptoms (nausea, vomiting, drowsiness, confusion, and fleeting numbness or tingling in the limbs). Hypertensive encephalopathy, a neurologic syndrome that occurs as a result of a sudden sustained rise in blood pressure, may be accompanied by nonspecific neurologic symptoms, such as confusion, headache, nausea, and even coma. It is also well recognized that end-stage renal disease (ESRD) is associated with accelerated and malignant hypertension; hypertension is associated with increased urinary calcium excretion and subsequent bone loss and osteoporosis, especially at the femoral neck.³⁸

MEDICAL MANAGEMENT

PREVENTION. The American Society of Hypertension recommends that everyone, regardless of age, know his or her blood pressure (the actual numbers). An annual blood pressure check is important for everyone; more frequent blood pressure measurements should be taken in anyone with risk factors or known hypertension. Elevated blood pressure in a younger adult (less than 50 years) can cause long-term accumulated damage, irreversible by age 50 or 60 years; therefore, any elevation in blood pressure at any age must be addressed.

The most important prevention factor is physical activity and exercise; other key variables include weight control, limitations on salt and alcohol intake, and modification of other risk factors present (see Box 12-8). Combining a low-sodium diet with the DASH (Dietary Approaches to Stop Hypertension) diet (high intake of fruits, vegetables, and low-fat dairy foods) helps reduce blood pressure more than the DASH diet alone, both for healthy people and for those with high blood pressure. Reducing salt intake also lowers the risk of osteoporosis and possible fracture, since a high salt intake increases urinary calcium excretion and hypertension has been found to be associated with bone loss.

DIAGNOSIS. In the past, hypertension was diagnosed primarily on the basis of the diastolic measurement, which was considered a better representation of the overall condition of the circulatory system. The rise of systolic pressure with age was considered to be normal and therefore

not a risk factor. Today, it is recognized that although diastolic hypertension (higher than 90 mm Hg) is common and usually controllable, systolic hypertension (higher than 140 mm Hg) is the most common in older adults and the most powerful risk factor for stroke and is strongly linked with heart attack, heart failure, and kidney failure even when the diastolic blood pressure is within normal limits. Systolic pressure measures the maximal strain on the heart and blood vessels and is a more precise measure of future damage to the system.

Blood pressure varies over the course of any single day depending on exertion, emotional state, ingestion of food, medications, and the presence of risk factors described previously. Thus it is important that blood pressure be measured at several different times and under consistent circumstances before a diagnosis of hypertension is made. Twenty-four-hour blood pressure monitoring using a portable device that automatically takes blood pressure readings at regular intervals is available and especially helpful in mapping out labile hypertension.

The individual maintains a log of activities and emotions corresponding to the times when readings are taken; this information is compared with the computer-generated map of blood pressures generated from the data collected by the measurement device. No other tests are specific for essential hypertension.

Studies used in the routine evaluation of hypertension may include a complete blood count (CBC); urinalysis; serum potassium, cholesterol, and creatinine assays; fasting blood glucose level; ECG; and chest radiography. Other more specific tests may be needed for secondary hypertension, and more complete cardiac assessment may be required for selected individuals.

TREATMENT. Once diagnosed, hypertension requires ongoing management (see Table 12-6) despite the absence of symptoms. According to the Joint National Committee,²⁶ the goal is to achieve and maintain the lowest safe arterial blood pressure (without side effects); the intended target goal is to reduce blood pressure to less than 135/85 mm Hg or 130/80 for people with diabetes or kidney disease.²⁶

The decision to treat and the method and intensity of intervention are based on the concept of total risk, not just blood pressure measurements. This approach takes into account cardiovascular risk factors in people with hypertension (see Box 12-8) and the presence of target organ damage or clinical cardiovascular disease (e.g., prior coronary revascularization, MI, stroke, PAD, retinopathy).

The WHO has published guidelines for the management of hypertension that review the management of risk factors in detail and prognosis based on risk stratification.³⁵⁸ In addition, a comprehensive list of recommendations on the treatment of hypertension in the prevention and management of ischemic heart disease is available from the American Heart Association.²⁸⁷

Management of hypertension may begin with a "stepped care" approach including smoking cessation and other nonpharmacologic interventions through lifestyle modification as initial therapy for primary hyperten-

sion (including those with blood pressure on the high side of normal or a family history of hypertension). This approach has been shown effective in lowering blood pressure and can reduce other cardiovascular risk factors.

Even when lifestyle modifications alone are not adequate in controlling hypertension, they may reduce the dosage of medication needed to manage the condition.⁷⁵ This program may include weight reduction; smoking cessation; a regular program of aerobic exercise; moderation of alcohol, dietary fat, caffeine, and dietary sodium; administration of nutritional supplements (e.g., potassium, calcium, magnesium); and behavioral cognitive therapy for those with hypertension associated with certain personality traits. See the previous section on Atherosclerosis: Treatment.

If nonpharmacologic measures fail to produce the desired results or if the blood pressure is very high at the time of diagnosis, blood pressure-lowering medications are prescribed starting with the lowest effective dose (to avoid intolerable side effects) and modifying accordingly. Antihypertensive medications can be classified by mode of action as diuretics, adrenergic blockers, vasodilators, ACE inhibitors, and calcium antagonists (see Table 12-5).

More than 50% of cases of mild hypertension can be controlled with one drug; a combination of medications may be required for others. Diuretics are often the first step in the pharmacologic management of hypertension. Although these drugs decrease plasma volume, potassium depletion and renal complications may require the use of β -blockers, calcium channel blockers, or ACE inhibitors.

Oral renin inhibitors such as aliskiren (Tekturna) are a new class of antihypertensive medication that can be used as a single treatment or in combination with other antihypertensive drugs. Aliskiren blocks the action of renin, a key kidney enzyme that helps regulate blood pressure. The blocking action takes place at the beginning of the renin-angiotensin-aldosterone system by directly inhibiting the renin released from the kidneys, thereby preventing conversion of angiotensinogen to angiotensin I. This is different from most other antihypertensive medications, which take action later in the renin-angiotensin-aldosterone system.

Black people are generally more responsive to calcium antagonists and diuretics than to β -blockers, ACE inhibitors, or the new oral renin inhibitors. Older adults with hypertension are generally equally responsive to all classes of antihypertensive medications, but they have an increased likelihood of side effects. Pharmacologic therapy is individualized, matching the individual's clinical presentation with medications available.

More aggressive early treatment of people with diabetes and elevated blood pressure is recommended. Hypertensive people who tend to be hostile may be told to take their medication at bedtime to avoid the sharp rises in blood pressure in the early morning hours associated with heart attacks. The use of home monitoring devices is an important part of the management program both to monitor the blood pressure and to evaluate the effect of antihypertensive medication.

Home monitoring can also distinguish between sustained hypertension and white coat hypertension and improves program compliance. In individuals with hypertension, the blood pressure readings taken in a clinic setting tend to be 5 to 10 mm Hg higher than measurements taken at home. Recommended frequency of readings is twice daily (morning and evening) on work and nonwork days for anyone newly diagnosed or in whom antihypertensive medication has recently been initiated or changed. Anyone with stable hypertension can take blood pressure reading several days per week.

Obesity has long been associated with hypertension and is an independent risk factor for cardiovascular disease and CAD as well. Regular exercise enhances weight loss and reduces blood pressure independent of weight loss. (For further discussion see Special Implications for the Therapist: Hypertension, section on exercise.)

Older individuals, blacks, and hypertensive individuals are more sensitive to change in dietary sodium chloride than are other individuals. A reduction of sodium intake alone may be enough to control blood pressure in persons with mild hypertension and may reduce the medication requirements in those who require drug therapy.

It is also recommended that individuals with high blood pressure limit their intake of alcohol to 2 oz of liquor, 10 oz of red wine, or 24 oz of beer per day. Women and lighter-weight men should limit alcohol intake to half of these amounts per day.⁷⁵

Dietary potassium deficiency may have a role in increasing blood pressure; individuals may also become hypokalemic from increased urinary magnesium excretion during diuretic therapy and require additional potassium. Magnesium and calcium influence vascular tone because magnesium acts to relax blood vessels and calcium assists in blood vessel contraction.

A proper balance of these two substances is essential, since they compete for entry into the cell. When magnesium is low, an abnormally large amount of calcium enters the cells so that blood vessels begin to lose their ability to relax. Progressive vasoconstriction and subsequent spasms result in elevated blood pressure and eventual ischemia. Muscle weakness with depressed tendon reflexes may accompany this condition. A fall in serum potassium level also enhances the effects of digitalis, increasing the risk of digoxin toxicity (see Table 12-5).

PROGNOSIS. Hypertension is a major risk factor for atherosclerosis, implicating hypertension as a common cause of death in the United States. Among black Americans, hypertension is also the most common fatal familial disease. More than one half of persons with angina pectoris, sudden death, stroke, and thrombotic occlusion of the abdominal aorta or its branches have hypertension.

Three fourths of people with CHF, dissecting aortic aneurysm, intracerebral hemorrhage, or rupture of the myocardial wall also have elevated blood pressure. If it is untreated, nearly 50% of people with hypertension die of heart disease, 33% die of stroke, and 10% to 15% die of renal failure.

When a person with hypertension achieves the target blood pressure, it must be emphasized that blood pressure control does not equal cure. Adherence to treatment and follow-up monitoring must be continued on an ongoing basis. Unfortunately, the cost of antihypertensives, side effects, and lack of symptoms sometimes lead to poor compliance with treatment. Treatment prolongs life, and antihypertensive medications have dramatically reduced the mortality rate associated with hypertension. See also the section on Atherosclerosis: Prognosis (especially regarding pulse pressure as the newest predictor of mortality).

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-4

Hypertension (Hypertensive Vascular Disease)

PREFERRED PRACTICE PATTERNS

4A: Primary Prevention/Risk Reduction for Skeletal Demineralization (osteoporosis prevention)

5A: Primary Prevention/Risk Reduction for Loss of Balance and Falling (Side Effect of Medication: Dizziness)

6A: Primary Prevention /Risk Reduction for Cardiovascular/Pulmonary Disorders

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

See also *Special Implications for the Therapist: Atherosclerosis*.

It is estimated that hypertension remains undiagnosed in nearly one half of the 60 million Americans who have it. It is possible that many people in a therapy practice will be hypertensive without knowing it. Cardiac pathology may be unknown, requiring the therapist to remain alert for risk factors that require medical screening. For anyone with identified risk factors, a baseline blood pressure measurement should be taken on two or three separate occasions, and any unusual findings should be reported to the physician. The role of the therapist in screening to identify conditions such as hypertension is important, since an essential early component of intervention for this condition includes exercise.

The potential for osteoporosis and subsequent hip fractures in older adults (especially women) with hypertension points to the importance of osteoporosis screening and prevention in this population. The physical therapist has an important role in the primary prevention of impairments and functional limitations in people with hypertension. A sudden increase in blood pressure such as occurs with any increase in intraabdominal pressure (e.g., Valsalva maneuver; see also Box 16-1) during exercise or stabilization exercises can be dangerous for already hypertensive persons. The therapist must alert individuals with hypertension to this effect and teach proper breathing techniques during all activities.

Medications

People with CAD taking NSAIDs for pain relief may also be at risk for a myocardial event during times of

increased myocardial oxygen demand (e.g., exercise, fever). In addition, older adults taking NSAIDs and antihypertensive agents must be monitored carefully. Regardless of the NSAID chosen, it is important to check blood pressure within the first few weeks after therapy or exercise is initiated and periodically thereafter.

A recent (2007) statement from the American Heart Association indicates that NSAIDs may cause an increased risk of serious cardiovascular thrombotic events, MI, stroke, heart failure, and hypertension.²⁵ Individuals with a prior history of cardiovascular disease or with risk factors for cardiovascular disease may be at greater risk. Specifically, a warning was issued for using selective inhibitors of cyclooxygenase-2 (COX-2).²⁵ Cyclooxygenase is an enzyme involved in production of prostaglandins, biologic mediators with a variety of functions. In the cardiovascular system, they regulate interactions between platelets and the vessel wall. As with all medications, a balance should be considered between the risks and benefits of NSAIDs.

Whenever a health care provider knows that a client has been prescribed antihypertensive medications, appropriate follow-up questions as to whether the client is taking the medication and taking it as prescribed must be addressed. Many people take the medication only when symptoms are perceived and are at risk for the complications described previously (especially during an exercise program, when oxygen demands of the myocardium increase).

Obtain as much information as possible about a client's medications so that potential side effects can be anticipated and intervention planned accordingly. Any side effects noted may indicate that a medication adjustment is needed and should be brought to the physician's attention (see Table 12-5).

The following brief description of the impact of various drug classes (all vasodilators) on exercise may assist the therapist in prescribing activities for those who require pharmacologic agents and provide insight into therapeutic decisions for active hypertensive individuals. Antihypertensive medications reduce resting blood pressure levels and may influence blood pressure changes during submaximal and maximal exertion, which affects exercise capacity.

Vasodilators such as nitroglycerin and other nitrates act as a prophylactic for angina by dilating the coronary arteries and improving collateral cardiac circulation, increasing oxygen to the heart muscle, and decreasing the blood pressure, thereby decreasing symptoms of angina.

The (3-adrenoceptor antagonists (β -blockers) selectively inhibit an increase in heart rate. Clinically, this means that when the person increases his or her activity or exercise level, the normal physiologic response of increased heart rate is blunted. This requires a longer warm-up and cool-down period. Sudden changes in position (e.g., supine to standing) should be avoided to prevent dizziness and falls associated with the resulting orthostatic hypotension (see

Continued.

Special Implications for the Therapist: Orthostatic Hypotension).

The β -blockers diminish catecholamine-induced elevations of heart rate, myocardial contractility, and blood pressure. These effects reduce myocardial oxygen requirements during exertion and stress, thereby preventing angina and allowing the person to exercise for longer periods before the onset of angina. The intended action of β -blockers may prevent normal blood pressure and heart rate responses to exercise; therefore, using heart rate as an index for monitoring response to exercise is not recommended. Although β -blockers are effective antihypertensives, most of them adversely alter aerobic capacity so that exercise capacity is reduced.

An exercise prescription should be based on exercise stress test results using recommended guidelines.⁹ Side effects of β -blockers include bronchospasm, which causes difficulty breathing, and chest tightness, which mimics angina; orthostatic hypotension; syncope; headache; and fatigue and weakness.

Diuretics have been first-line antihypertensive agents for many years, but few studies have observed the effect of diuretic therapy on exercise performance. Existing evidence reveals that peak blood pressures induced by physical activity may not always be controlled with diuretics. Diuretic therapy can result in hypokalemia accompanied by muscular cramps and skeletal muscle fatigue (see also Chapter 5).

Potassium-sparing diuretics may cause hyperkalemia, which can in turn cause ventricular arrhythmias. Exercise tolerance may be reduced with arrhythmias because of a decrease in left ventricular filling time. Prolonged exercise in the heat is not recommended for people taking diuretics because of the cumulative effects of heat, exercise, and diuretics on blood volume and electrolytes. The length of time an individual who is taking a diuretic can safely exercise in the heat varies with the heat index and the physical condition of the person.

Calcium plays a role in the electrical excitation of cardiac cells and in the mechanical contraction of the myocardial and vascular smooth muscle cells. Calcium channel antagonists inhibit calcium ion influx across the cell membrane during cardiac depolarization, relax coronary vascular smooth muscle, dilate coronary and peripheral arteries, and increase myocardial oxygen delivery in people with vasospastic angina. This class of vasodilators decreases peripheral vascular resistance at rest and during physical activity, thereby altering exercise tolerance by affecting heart rate and blood pressure during exercise.

During exercise, calcium channel antagonists have been observed to reduce systolic and diastolic pressure at submaximal loads, but higher systolic blood pressures measured during maximal exercise are not lowered. Side effects of calcium antagonists (e.g., drowsiness, dizziness, headache, peripheral edema, tachycardia or bradycardia) may interfere with a client's ability to participate in an exercise program.

ACE inhibitors reduce blood pressure by lowering peripheral vascular resistance and are considered the

first line of treatment by many physicians. They are readily available as generic drugs (less expensive). Contrary to expectation, the aerobic exercise capacity of patients was found to be greater with the lower dose of lisinopril, suggesting that therapy with ACE inhibitors for heart failure may require tailoring the doses to the individual to optimize functional benefits.⁸⁴

Use of calcium channel antagonists and ACE inhibitors in hypertension increased dramatically in the 1990s, whereas the use of less expensive agents, such as diuretics and β -blockers, declined.

Exercise and Blood Pressure

The benefits of resistive or dynamic exercise in people with and without cardiovascular disease are well known and available for review.²⁶⁸ A regular program of aerobic exercise, introduced gradually, facilitates cardiovascular conditioning, may assist in weight reduction, and may provide some benefit in reducing blood pressure. Exercising using primarily the lower extremities (e.g., cycling, walking) can also reduce blood pressure.

Postexercise hypotension (lowered blood pressure in response to exercise) in mildly hypertensive individuals has been observed for up to 7 hours after exercise independent of other variables.²⁶⁴ Diastolic blood pressure reduction seems to be related to the duration of the exercise program (i.e., the longer the program, the more likely the hypotensive effect). Blood pressure reduction has occurred after just several weeks to 6 months of regular training.¹⁴⁰ On the other hand, blood pressure will return to its previous elevated level if training is discontinued.

Heavy isometric exercises and heavy weightlifting may be harmful, since the blood pressure often rises because of vasovagal reflexes that occur. During fatiguing isometric exercise, the rate and rise of systolic blood pressure appear to be higher in hypertensive individuals, but studies in this area are limited. Generally, antihypertensive drugs have not been found to affect the blood pressure response to isometric exertion. However, the use of isometric exercise to lower blood pressure has not been studied in hypertensive individuals; a fall in resting blood pressure has been observed in normotensive individuals after repetitive isometric contractions equal to 30% of maximal capacity.³⁵⁹

Exercise Training Guidelines

The intensity of exercise required to produce health benefits and decrease blood pressure has been confused with the level of exercise necessary to improve physical fitness. Health benefits can be achieved without large gains in fitness. Encouraging people to increase their level of total energy expenditure is the key to increasing activity levels, rather than emphasizing physical fitness. The type, intensity, duration, and frequency of training, as well as progression, should be assessed regularly.

A preexercise evaluation and exercise testing may be prescribed by the physician. This information is helpful

in establishing submaximal and maximal blood pressure responses. Monitoring vital signs before, during, and after exercise or activity is essential. Any person with an exaggerated systolic blood pressure response (higher than 250 mm Hg) or failure to reduce diastolic pressure (to less than 90 mm Hg) should be referred to the physician for reevaluation.

Training intensity does not need to be high, and it appears that low-intensity activity (65% to 70% of maximal heart rate) three times per week is as effective as high-intensity activity in blood pressure reduction. Training intensity should be based on maximal heart rate using the calculated formulas (see Appendix B) or measured during a maximal exercise test. After 12 to 16 weeks, if the blood pressure is adequately controlled, the physician may reduce the antihypertensive medication slowly to determine the long-term effect of training on blood pressure. Several resources are available for determining the appropriate exercise program for the hypertensive client, whether symptomatic or asymptomatic.^{9,44}

Monitoring during Exercise

Therapists often treat people who are diagnosed with conditions that are highly correlated with hypertension, such as stroke, obesity, diabetes mellitus, alcoholism, CAD, and pregnancy (see Box 12-7). Monitoring tolerance to exercise by observing for unusual symptoms and measuring blood pressure before, during, and after therapy are important steps in identifying a potential cardiovascular event. Many factors can cause an increase in blood pressure (see Appendix B).

Hypertensive Heart Disease

Definition and Overview. The term *hypertensive heart disease* is used when the heart is enlarged as a result of persistently elevated blood pressure (hypertension) (see previous discussion). Left ventricular hypertrophy and diastolic dysfunction are found in 10% to 30% of the adult population with chronic hypertension, and it may present with many of the signs and symptoms of CHF. Both the prevalence and the severity of the disease are greater in blacks than in whites. In all adults, it increases progressively with age.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Cardiac enlargement and left ventricular hypertrophy, best viewed with echocardiography, are diagnostic of hypertensive heart disease. Treatment is as for hypertension unless heart failure develops, in which case treatment is as for heart failure (see the sections on each of these conditions for more discussion). The most common cause of death in hypertensive heart disease is CHF, accounting for 40% of all deaths from hypertension.

SPECIAL IMPLICATIONS FOR THE THERAPIST

12-5

Hypertensive Heart Disease

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

See Special Implications for the Therapist: Atherosclerosis, Special Implications for the Therapist: Hypertension, and Special Implications for the Therapist: Congestive Heart Failure.

Myocardial Infarction

Definition and Incidence

MI, also known as a heart attack or referred to as a "coronary," is the development of ischemia with resultant necrosis of myocardial tissue. Any prolonged obstruction depriving the heart muscle of oxygen can cause an MI. MI occurs in 1 $\frac{1}{2}$ million persons each year and represents the leading cause of death (500,000 deaths annually) in the adult American population. See the previous section on Ischemic Heart Disease in this chapter.

Etiologic and Risk Factors

Etiologic and risk factors are the same as for all forms of cardiovascular disease, especially angina pectoris associated with CAD (see the previous section on Angina Pectoris). Eighty percent to ninety percent of MIs result from coronary thrombus at the site of a preexisting atherosclerotic stenosis. New cases of MI occur in many people with only a borderline risk profile or even lack of known risk factors, suggesting other unidentified risk factors.

Other causes may include cocaine use (causes vasoconstriction of the coronary arteries), vasculitis, aortic stenosis, or aortic root or coronary artery dissection. Smokers have more than twice as many heart attacks as nonsmokers, and sudden cardiac death occurs two to four times more frequently in smokers. After an infarction, smokers have a poorer chance of recovery than nonsmokers. Exertion-related MI, which may include weakness or shortness of breath while working with the arms extended overhead in habitually inactive people, has also been reported associated with single-vessel rather than triple-vessel disease.¹²⁶

It is a well-established fact that heart attacks occur more frequently in the early morning hours. This peak incidence is attributed to an increase in catecholamines with the resultant increased blood pressure, increased workload of the heart, as well as increased clotting factors in the early morning. Heart attacks also occur in a seasonal pattern with an increased incidence between Thanksgiving and New Year's Day across all ages, in both genders, and across geographic regions. Whether this can be attributed to mood changes, weather, circadian rhythms, large quantity of food consumed, or some other mechanism remains unknown.

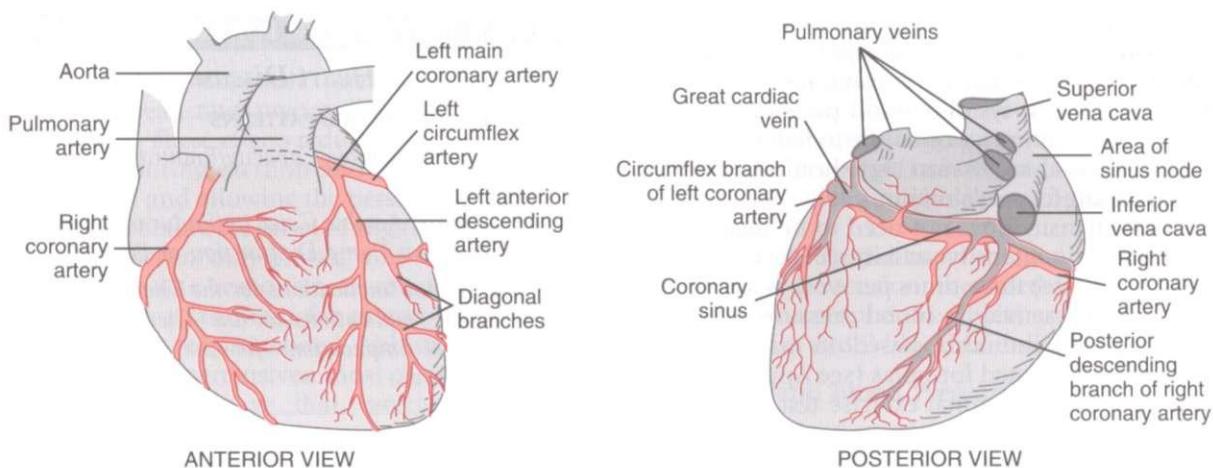


Figure 12-8

Areas of myocardium affected by arterial insufficiency of specific coronary arteries. The right and left coronary arteries branch off the aorta just above the aortic valve and normally supply the myocardium with oxygenated blood. The most commonly affected arteries and the area of myocardium supplied are listed in Table 12-9.

Upper respiratory tract illnesses have been associated with an increased risk of ischemic heart disease and stroke, especially during the flu season in adults 65 years old and older who have not received a flu shot. Studies show a reduction in the risk of hospitalization and mortality for heart disease as well as cerebrovascular disease, pneumonia, and influenza among elderly adults receiving flu vaccine.^{240,241}

The association between periodontal disease (e.g., gingivitis, periodontitis) and acute MI is under investigation. There is a definite association between common forms of periodontal disease and cardiovascular disease and stroke, but the causal relations have not been identified.²⁴⁵

Researchers have found that bacteria in the mouth spill into the bloodstream and can be found in the walls of major arteries. Recent research showed that intensive periodontal treatment may reverse atherosclerosis by improving elasticity of the arteries, or endothelial function,³³³ suggesting that periodontal treatment may reduce cardiovascular risk.

Acute respiratory tract infections, such as the common cold, flu, and bronchitis, may also increase the risk of a heart attack's occurring within 2 weeks of a first heart attack and may account for some cases of MI, although researchers point out that early symptoms of heart attacks may be mistaken for acute upper respiratory tract infection.²¹⁷

Silent ischemia is highly prevalent among people with diabetes; increased PAI-1 activity has been identified as a risk factor for MI in persons with diabetes as well as for postmenopausal women not receiving hormone replacement therapy. PAI-1 is a naturally occurring substance that inhibits another natural substance, t-PA; t-PA is an enzyme released endogenously as part of the body's defense against thrombosis; it lyses fibrin and dissolves the forming blood clot (thrombus). The effect of PAI-1 on t-PA is to prevent clot destruction in the bloodstream.

Research now shows that diabetic clients have higher PAI-1 activity than nondiabetic clients, both at hospital

admission for acute MI and at follow-up 1 year later. Raised PAI-1 activity may predispose diabetic clients to MI and may also impair pharmacologic and spontaneous reperfusion after acute MI, contributing to the poor outcome in this population.³²⁴

Pathogenesis

The myocardium receives its blood supply from the two large coronary arteries and their branches (Fig. 12-8; Table 12-9). One or more of these blood vessels may become occluded by a clot that forms suddenly when an atheromatous plaque ruptures through the sublayers of a blood vessel or when the narrow, roughened inner lining of a sclerosed artery becomes completely filled with thrombus. In most cases, infarcts result from an occlusive thrombus superimposed on an atherosclerotic plaque.

Researchers have found that plaque most likely to rupture (vulnerable plaque) is comprised of the soft form of cholesterol (cholesteryl ester) and is vulnerable to mechanical forces such as occur with the increase in hormones early in the morning or even the vibration of the heartbeat.

Rupturing plaque does not always result in an MI. It is likely that plaque breaks off frequently without triggering a heart attack, and the large plaques visible on angiograms are often the healed-over and more stable plaques. Although these plaques occlude the coronary vessels, resulting in obstruction, ischemia, and angina, they are not as likely to cause rupture and sudden death as happens with the soft, smaller, and usually undetected plaques.

The most common site involved is the left ventricle, the chamber of the heart with the greatest workload. Thrombosis of the anterior descending branch of the left coronary artery is the most common cause of infarction and affects the anterior left ventricle (Fig. 12-9).

Occlusion of the left circumflex artery produces anterolateral or posterolateral infarction. Right coronary artery thrombosis leads to infarction of the posteroanterior portion of the left ventricle and may involve the right ventricular myocardium and interventricular septum. The

arteries supplying the atrioventricular node and the sinus node more commonly arise from the right coronary artery; thus atrioventricular block at the nodal level and sinus node dysfunction occur more frequently during inferior infarctions.

Myocardial ischemia/reperfusion injury is accompanied by an inflammatory response involving three major components: (1) molecular oxygen, (2) cellular blood elements (especially neutrophils), and (3) activated complement system. When the myocardium has been completely deprived of oxygen, cells die and the tissue becomes necrotic in an area called the *zone of infarction* (Fig. 12-10; see also Figs. 6-5 and 6-6). In response to this necrosis, leukocytes aid in removing the dead cells, and fibroblasts form a connective tissue scar within the area of infarction. The remaining heart muscle cells enlarge to

compensate for the loss in heart pump function (see also the section on Cell Injury in Chapter 6; a complete discussion of the role of oxidative stress and complement activation in heart disease is available^{71,99}). Usually the formation of fibrous scar tissue is complete within 6 to 8 weeks (Table 12-10; see Fig. 6-6).

Immediately surrounding the area of infarction is a less seriously damaged area of injury called the *zone of hypoxic injury*. This zone is able to return to normal, but it may also become necrotic if blood flow is not restored. With adequate collateral circulation, this area may regain its function within 2 to 3 weeks. Adjacent to the zone of

Table 12-9 Blood Supply to the Myocardium

Area of Myocardium*	Supplied by
Anterior	Left coronary artery Left anterior descending branch†
Posterior	Right coronary artery
Inferior	Right coronary artery
Anteroseptal	Left coronary artery Left anterior descending branch
High lateral	Circumflex artery Left coronary artery Diagonal branch
Apical	Usually left coronary artery Left anterior branch Sometimes right coronary artery Posterior descending branch

*Most commonly affected arteries and the area of myocardium supplied are listed in order of decreasing frequency of blockage.

†Often referred to as the "widow maker," untreated blockage of the left anterior descending branch leads to permanent heart damage if the individual does not die first.

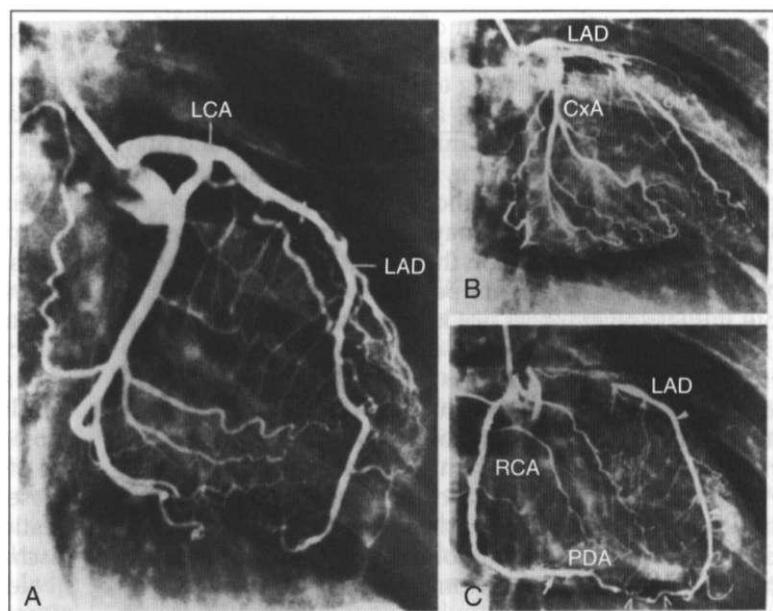
Table 12-10 Tissue Changes After Myocardial Infarction (MI)

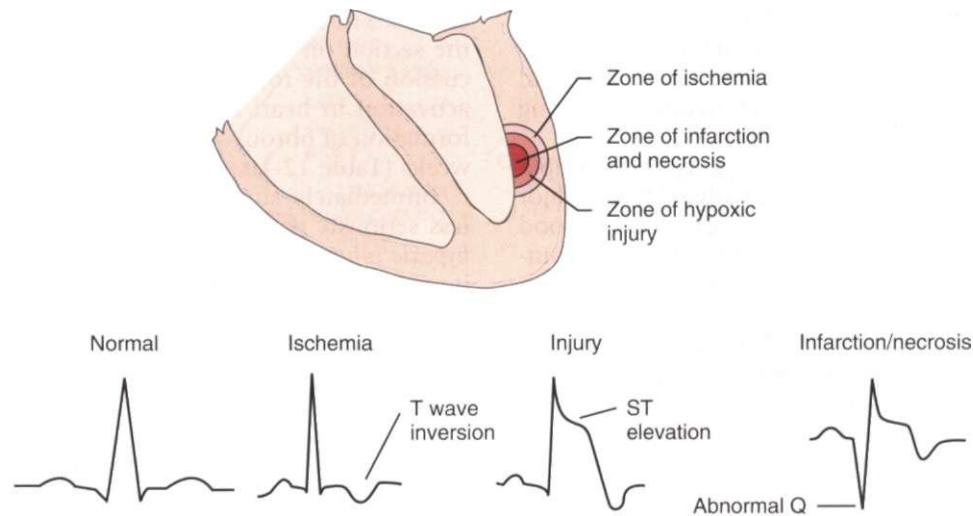
Time after MI	Tissue Changes
6-12 hr	No gross changes; healing process has not begun
18-24 hr	Inflammatory response; intercellular enzyme release
2-4 days	Visible necrosis; proteolytic enzymes remove debris; catecholamines, lipolysis, and glycogenolysis elevate plasma glucose and increase free fatty acids to assist depleted myocardium recovery from anaerobic state
4-10 days	Debris cleared; collagen matrix laid down
10-14 days	Weak, fibrotic scar tissue with beginning revascularization; area vulnerable to stress
6 wk	Scarring usually complete; tough, inelastic scar replaces necrotic myocardium; unable to contract and relax like healthy myocardial tissue

Modified from McCance KL, Huether SE: *Pathophysiology: the biologic basis for disease in adults and children*, ed 5, St Louis, 2006, Mosby.

Figure 12-9

A, Angiogram of a normal left coronary artery (LCA). **B**, Angiogram of a totally obstructed left anterior descending (LAD) coronary artery. **C**, Angiogram of the right coronary artery (RCA) and its major branch, the posterior descending artery (PDA) (same heart as in **B**). The LAD is seen because of collateral vessels connecting the LAD and the RCA system. (From Boucek R, Morales A, Romanello R, et al: *Coronary artery disease: pathologic and clinical assessment*, Baltimore, 1984, Williams & Wilkins, pp 4, 9.)



**Figure 12-10**

Electrocardiographic (ECG) alterations associated with the three zones of myocardial infarction (MI).

hypoxic injury is another reversible zone called the *zone of ischemia*. Ischemic and injured myocardial tissues cause characteristic ECG changes; as the myocardium heals, the ST segment and T waves gradually return to normal, but abnormal Q waves may persist.

Oxygen deprivation is accompanied by electrolyte disturbances, particularly cellular loss of potassium, calcium, and magnesium. Myocardial cells deprived of necessary oxygen and nutrients lose contractility, thereby diminishing the pumping ability of the heart. Normally the myocardium takes up varying quantities of catecholamines (epinephrine, norepinephrine), which are released when significant arterial occlusion occurs. Released catecholamines predispose the individual to serious imbalances of sympathetic and parasympathetic function, irregular heartbeats (arrhythmia), and heart failure.

Clinical Manifestations

The most notable symptom of MI is a sudden sensation of pressure, often described as prolonged crushing chest pain, occasionally radiating to the arms, throat, neck (as high as the occipital area), and back (Fig. 12-11). The pain is constant, lasting 30 minutes up to hours, and may be accompanied by pallor, shortness of breath, and profuse perspiration. Catecholamine release resulting in sympathetic stimulation may produce diaphoresis and peripheral vasoconstriction that cause the skin to become cool and clammy. Angina pectoris pain can be similar, but it is less severe, does not last for hours, and is relieved by cessation of activity, rest, or nitrates.

Symptoms do not always follow the classic pattern, especially in women. Two major symptoms in women are shortness of breath, sometimes occurring in the middle of the night, and chronic, unexplained fatigue. Atypical presentation may include continuous pain in the midthoracic spine or interscapular area, neck and shoulder pain, stomach or abdominal pain, nausea, unexplained anxiety, or heartburn that is not altered by antacids.

Silent attacks (painless infarction without acute symptoms) are more common among nonwhites, older adults (more than 75 years), all smokers, and adults (men and women) with diabetes, presumably because of reduced sensitivity to pain. Nausea and vomiting may occur because of reflex stimulation of vomiting centers by pain fibers. Fever may develop in the first 24 hours and persist for a week because of inflammatory activity within the myocardium.

Postinfarction complications include arrhythmias, CHF, cardiogenic shock, pericarditis, rupture of the heart, thromboembolism, recurrent infarction, and sudden death. Arrhythmias, affecting more than 90% of individuals, are the most common complication of acute MI and are caused by ischemia, hypoxia, autonomic nervous system imbalances, lactic acidosis, electrolyte imbalances, drug toxicity, or alterations of impulse conduction pathways or conduction defects.

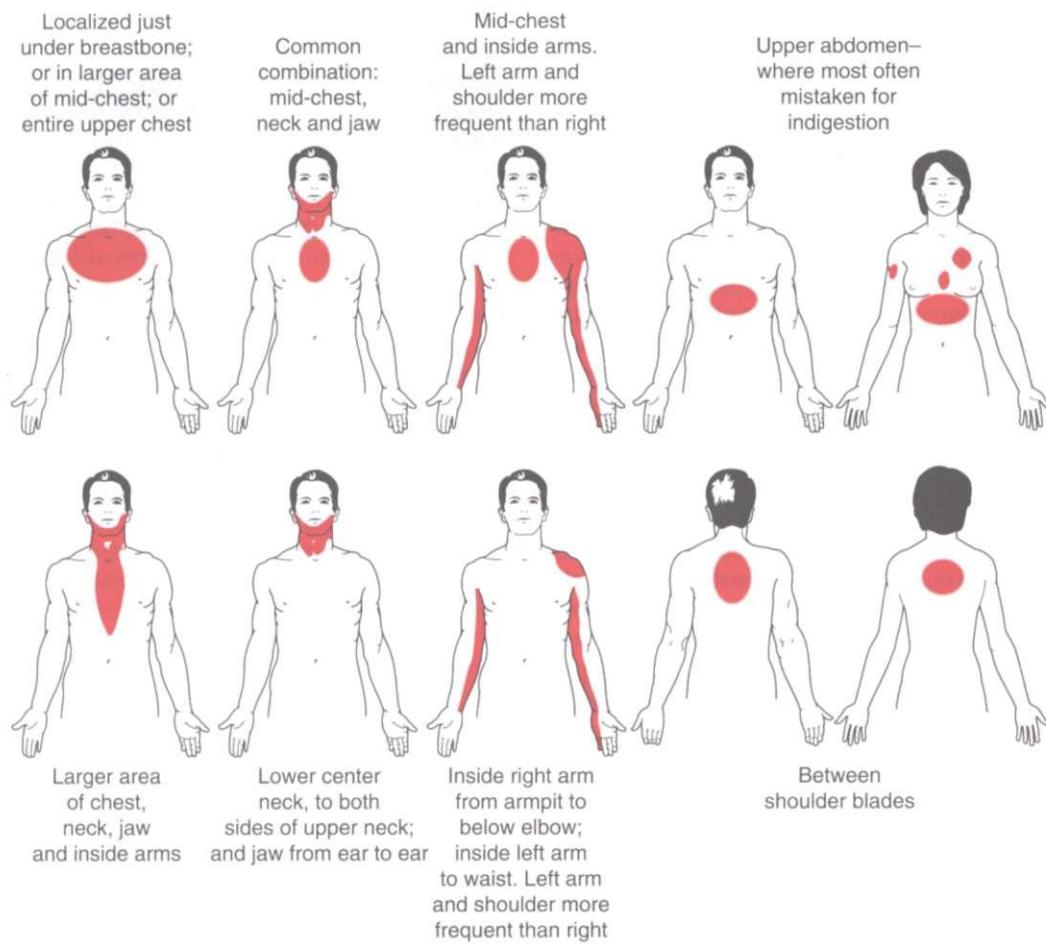
MEDICAL MANAGEMENT

PREVENTION. See the sections on Atherosclerosis; Prevention and Angina Pectoris: Prevention and Treatment.

DIAGNOSIS. Diagnosis of acute MI and determination of the site and extent of necrosis rely on the clinical history, interpretation of the ECG, and measurement of serum levels of cardiac enzymes. Diagnostic uncertainty frequently arises because of a variety of factors.

Many people with acute MI have atypical symptoms, and one half of all people with typical symptoms do not have acute MI. One half of the people with acute MI have nondiagnostic ECGs, and some people are unable to provide a history. Biochemical markers of cardiac injury are commonly relied on to diagnose or exclude acute MI. These laboratory tests dramatically reduce the cost of treating heart attacks by allowing physicians to quickly discharge people with noncardiac chest pain.

Newer biochemical markers of myocardial injury, such as cardiac troponin I (TnI) and cardiac troponin T (TnT)

**Most common warning signs of heart attack**

- Uncomfortable pressure, fullness, squeezing or pain in the center of the chest (prolonged)
- Pain that spreads to the throat, neck, back, jaw, shoulders, or arms
- Chest discomfort with lightheadedness, dizziness, sweating, pallor, nausea, or shortness of breath
- Prolonged symptoms unrelieved by antacids, nitroglycerin, or rest

Atypical, less common warning signs (especially women)

- Unusual chest pain (quality, location, e.g., burning, heaviness; left chest), stomach or abdominal pain
- Continuous midthoracic or interscapular pain
- Continuous neck or shoulder pain
- Isolated right biceps pain
- Pain relieved by antacids; pain unrelieved by rest or nitroglycerin
- Nausea and vomiting; flu-like manifestation without chest pain/discomfort
- Unexplained intense anxiety, weakness, or fatigue
- Breathlessness, dizziness

Figure 12-11

Early warning signs of a heart attack. Multiple segmental nerve innervation shown in Fig. 12-6 accounts for the varied pain patterns possible. A woman can experience any of the various patterns described but is more likely to develop atypical symptoms of pain as depicted here. (Modified from Goodman CC, Snyder TE: *Differential diagnosis for physical therapists: screening for referral*, ed 4, Philadelphia, 2007, Saunders.)

(regulatory proteins that help the heart muscle contract), are now being used instead of or along with the standard markers, such as the myocardial isoenzyme of creatine kinase (CK-MB). TnT is quite specific for myocardial ischemia and necrosis. It remains elevated 5 to 7 days after an MI and is a predictor of cardiovascular mortality.

TnI is a better cardiac marker than CK-MB for MI because it is more sensitive and more specific to myocardial injury; TnT is a better predictor of cardiovascular mortality (as well as all-cause mortality). Both TnI and TnT are useful markers for myocardial injury that help determine the prognosis in people who have unstable

angina but no evidence of CK-MB elevation. (See also the section on Cardiac Enzymes and Markers in Chapter 40; see Tables 40-15 and 40-16.)

Researchers are continuing to investigate other hemostatic markers based on the knowledge that coronary thrombosis involves both coagulation and fibrinolysis cascades. For example, increases of fibrinogen and D-dimer, a circulating marker of fibrin turnover, are significantly higher in people with acute ischemic events such as MI and unstable angina than in nonischemic individuals, but it has not been determined to what extent this is causal. Other tests may include nuclear scanning, coro-

nary angiography, echocardiography, CT, cardiac magnetic resonance (CMR) stress testing, and MRA. Serum cholesterol must be determined because of its importance as a modifiable risk factor. See the previous sections on Angina Pectoris: Diagnosis and Atherosclerosis: Diagnosis.

Other cardiac markers include homocysteine, Lp(a), and CRP. Although these have not become "standard" laboratory values, they can be used as independent predictors of future coronary events in apparently healthy men and women. For example, elevated plasma total homocysteine is a risk factor for atherosclerosis and endothelial dysfunction, and CRP may be used as a marker of subclinical atherosclerosis and cardiovascular risk specifically linked to MI and sudden death.

Infarcted tissue is electrically silent and does not contribute to the ECG. Most clients with acute infarction have ECG changes, although this test provides only a crude estimate of the magnitude of infarction. When diagnosis by ECG and enzymes is not possible (e.g., when people seek medical attention after MI), scintigraphic studies (radionuclide imaging) can show areas of necrotic myocardium and diminished perfusion. These tests, which use radiotracers, do not distinguish old damage from recent infarction, and false-positive results can occur.

Other test procedures may include echocardiography, which is useful in assessing the ability of the heart walls to contract and relax, and transesophageal echocardiography (TEE), an ultrasonic technique that provides a clearer image of the heart, including the posterior wall, valvular anatomy, and thoracic aortic structure, providing identification of structural heart diseases. Newer technology, such as RT-3D imaging, has the potential to improve evaluation of heart function (especially ventricular) with TEE.

Magnetic resonance imaging (MRI) to evaluate structural defects of the heart and positron emission tomography (PET) to evaluate cardiac physiology and metabolism and assess tissue perfusion have contributed significantly to the understanding of the pathophysiology of the ischemic heart.

Another new technique being investigated is the use of a contrast agent called EchoGen, used in conjunction with an ultrasound procedure. This agent infiltrates healthy heart muscle but not muscle that has been deprived of blood or oxygen. Existing contrast agents only image the heart chambers, which provides information about the flow of blood through the chamber but not about the structure of the heart muscle itself.

TREATMENT. The goal of treatment is reestablishing the flow of blood in blocked coronary arteries. Pharmacologic intervention is used to provide pain relief (essential since angina is evidence of ongoing ischemia), limit infarction size, reduce vasoconstriction, prevent thrombus formation, and augment repair. MI caused by intracoronary thrombi can be relieved by infusion of thrombolytic agents (e.g., streptokinase, urokinase, t-PA) that dissolve clots, promote vasodilation, and reduce infarct size.

PAI-1 is a naturally occurring substance that inhibits another natural substance, t-PA; t-PA is an enzyme released endogenously as part of the body's defense against thrombosis; it promotes degradation of fibrin leading to dissolving of blood clots. The effect of PAI-1 on t-PA is to *prevent* clot destruction in the bloodstream.

Tissue plasminogen activator, a naturally occurring enzyme that promotes dissolving of blood clots, is now a genetically engineered drug used in thrombolytic therapy. However, a single dose of recombinant t-PA (rt-PA) costs about \$1000, whereas other drugs are less expensive (e.g., streptokinase costs about \$300).

This intervention initiated within 70 minutes of symptom onset is associated with improved outcome.³⁴⁸ After a thrombolytic agent is administered, intravenous (IV) heparin therapy is usually given with adjunctive drug therapy during and after MI, because platelet inhibitors and other cardiovascular medications (see Table 12-5) are known to further reduce mortality when administered during the acute phase.

Right now, only 5% of heart attack victims receive reperfusion therapy within that crucial first hour after symptom onset, primarily because people delay (sometimes by hours) coming to the emergency department. This points out the extreme importance of early intervention and education of the general population (and especially for those with known risk factors, such as hypertension, previous heart attack, diabetes, smoking, or hyperlipidemia) as to the importance of getting to an emergency department at the earliest sign of heart attack. Educating the public about the less common or atypical warning signs and symptoms is essential. Information about public education, reducing delays at home or at work, and the National Heart Attack Alert Program is available.^{88,237}

Other treatment interventions, including identification and modification of risk factors, angioplasty, stenting, atherectomy, angiogenesis, tissue engineering, gene therapy, stem cell transplantation, and cardiac rehabilitation utilizing exercise programs, have been previously discussed in detail (see the sections on Atherosclerosis: Medical Management and Hypertension: Medical Management).

A study to determine whether early, rapid use of cholesterol-lowering therapy can reduce recurrent ischemic events in acute coronary syndromes is under way through the MIRACL (Myocardial Ischemia Reduction with Aggressive Cholesterol Lowering) program. The study showed that lipid-lowering therapy with 80 mg/day of atorvastatin, initiated during acute coronary syndrome, reduces recurrent ischemic events in patients in the first 16 weeks.^{255,364}

Exercise has been recommended as a means of increasing pain tolerance, increasing the threshold of the stimulus required to induce angina, alleviating depression, reducing anxiety, and inducing collateral circulation. Increasing evidence suggests that combining a low-fat diet and intensive exercise training can improve myocardial perfusion by regression of coronary atherosclerosis. Exercise training may be contraindicated for some people

Box 12-9**CONTRAINDICATIONS TO EXERCISE AFTER MYOCARDIAL INFARCTION (MI)**

- Acute MI (<1 or 2 days after an MI without physician approval)
- Unstable angina; easily provoked angina
- New electrocardiogram (ECG) with abnormalities
- Signs and symptoms of MI (e.g., nausea, dyspnea, light-headedness, chest pain)
- Arterial partial pressure of oxygen (PaO_2) <60 mm Hg
- O_2 saturation <85%
- Hemoglobin <8 g/dl; hematocrit <26%
- Severe aortic outflow obstruction
- Suspected or known dissecting aneurysm
- Acute myocarditis or pericarditis
- Uncontrolled complex arrhythmias
- Active severe congestive heart failure (CHF); resting respiratory rate >45 breaths/min
- Recent pulmonary embolism or thrombophlebitis
- Untreated third-degree heart block
- Severe systemic hypertension unresponsive to medication
- Uncontrolled diabetes
- Acute infections
- Digoxin toxicity (see Table 12-5)

Modified from Kavanagh T: Cardiac rehabilitation. In Goldberg L, Elliott DL, eds: *Exercise for prevention and treatment of illness*, Philadelphia, 1994, FA Davis, p 55.

(Box 12-9; see also Box 12-4). Medical clearance must be obtained for entry into an exercise training program.

Exercise testing is the most useful tool to establish guidelines for exercise training in apparently healthy adults and is mandatory for people with known or suspected cardiovascular disease.^{108,109} The majority of exercise testing can be done within 3 days of MI with a very low incidence of complications. Criteria for testing usually include clients who are off IV nitroglycerin with no angina at rest, uncontrolled cardiac failure, or arrhythmias. Early testing can lead to early triage and potential cost savings.

PROGNOSIS. The size and anatomic location of the infarction, together with the amount of damage from previous infarctions, determine the acute clinical picture, the early complications, and the long-term prognosis. The first 24 hours after onset of symptoms is the time of highest risk for sudden death. The sooner someone reaches the hospital, the better the prognosis. Eighty percent of those experiencing an acute MI survive the initial attack when transported to a coronary care unit (CCU). Substantial reductions in post-MI death have occurred over the last five decades because of improved intervention.

Factors negatively affecting prognosis include age (clients older than 80 years have a 60% mortality); evidence of other cardiovascular diseases, respiratory diseases, or uncontrolled diabetes mellitus; anterior location of MI (30% mortality rate); and hypotension (clients whose systolic blood pressure is less than 55 mm Hg have a 60% mortality rate). The risk of reinfarction is increased in women, people with elevated blood pres-

sure, and people with elevated serum cholesterol. As MI survivors with long-standing hypertension live longer, cardiac failure has become an increasingly important long-term sequela of MI.

Prognostic testing predictive of cardiac events includes standard exercise testing such as functional capacity and heart rate recovery¹⁰⁸ and imaging using SPECT with contrast agents (e.g., thallium Tl 201, technetium Tc 99m sestamibi). In the imaging studies, a radioisotope is taken up by adequately perfused tissue, allowing detection of myocardial perfusion defects at rest and during exercise (areas of infarction appear as regions of diminished isotope activity or no activity, referred to as *cold spots*).

Study of the prognostic value of treadmill exercise testing in older persons has shown that workload (measured in metabolic equivalents) is the only treadmill exercise testing predictive of death both in younger persons and in adults over 65 years of age.¹³⁰ An abnormal exercise test result is a more powerful predictor of risk in those people with conventional risk factors than in those without such risk factors.

SPECIAL IMPLICATIONS FOR THE THERAPIST**12-6*****Myocardial Infarction*****PREFERRED PRACTICE PATTERNS**

6A: Primary Prevention/Risk Reduction for Cardiovascular/Pulmonary Disorders

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

Other patterns may apply depending on postinfarction complications (e.g., arrhythmias, CHF, embolism, side effects of medications). See also Special Implications for the Therapist: Atherosclerosis.

Early Post-Myocardial Infarction Considerations

Although the myocardium must rest, bed rest puts the client at risk for development of hypovolemia (low blood volume), hypoxemia (hypoxia), muscle atrophy, and pulmonary embolus (PE; see also the section on The Cardiac Client and Surgery in this chapter). Developing a program of progressive physical activity with adequate pacing and rest periods begins within 24 hours for the acute care client in uncomplicated cases.

Gentle movement exercises, deep breathing, and coughing are usually begun immediately as prophylactic measures. Incisional pain or discomfort from cardiac surgery may cause a person to exhibit rapid, shallow respirations in an attempt to ease the discomfort. If analgesics are prescribed to prevent severe discomfort, the drug can be administered before therapy to better enable the person to carry out breathing exercises. This problem is of limited duration and usually resolves when the incision heals. The therapist must be aware that analgesics also mask pain response, making it possible for the client to overexert.

Continued.

Early therapeutic exercise helps prevent cardiopulmonary complications, venous stasis, joint stiffness, and muscle weakness. Relaxation is often promoted with low-intensity activity. Activities that increase intrathoracic or intraabdominal pressure, such as breath holding and Valsalva maneuvers (see Box 16-1), can precipitate bradycardia followed by increased venous return to the heart, causing possible cardiac overload. For this reason, these actions are contraindicated and should not be performed in any stage of the rehabilitation program.⁹²

During the first 6 weeks post-MI, the client is cautioned to avoid saunas, hot tubs, whirlpools, and excessively warm swimming pools. Early rehabilitation lasting 2 to 3 weeks is often followed by exercise testing, at which time water therapy may be permissible per physician approval (see Guidelines for Aquatic Therapy in Appendix B). Specific aspects of cardiac rehabilitation and postoperative care are beyond the scope of this text. Other more specific texts are available to guide the therapist in this area.^{114,150,162} See also the Agency for Health Care Policy and Research's clinical guidelines for cardiac rehabilitation.⁴

Monitoring Vital Signs

The therapist must continually monitor for signs of impending infarction, including generalized or localized pain anywhere over the thorax, upper limbs, and neck; palpitations; dyspnea; light-headedness; syncope; sensation of indigestion; hiccups; and nausea (see Fig. 12-11). Pain medications, such as morphine, used to minimize discomfort initially may also depress the respiratory drive.

The CCU therapist must monitor corresponding vital signs. The home health therapist must monitor pulse and blood pressure measurements for hypotension because of the side effects precipitated by antihypertensive medications, vasodilators, and other antianginal agents. (See also Special Implications for the Therapist: Angina Pectoris.) Initial ambulation and activities at home should be roughly equivalent to levels achieved at the hospital at the time of discharge, depending on the client's physiologic response to the transition from hospital to home.

The client must increase activities gradually to avoid overtaxing the heart as it pumps oxygenated blood to the muscles. The metabolic equivalent system provides one way of measuring the amount of oxygen needed to perform an activity: 1 metabolic equivalent of the task (MET) equals 3.5 ml of oxygen per kilogram of body weight per minute; 1 MET is approximately equivalent to the oxygen uptake a person requires when resting. At 2 METs, the individual is working at twice his or her resting metabolic rate.

Early mobilization activities after acute MI should not exceed 1 to 2 METs (e.g., brushing teeth, eating). By comparison, people who can exercise to 8 or more METs (oxygen uptake of 28 ml/kg/min or more) can perform most daily physical activities. In general, 3 to 6 METs is considered the equivalent of moderate exercise. Activities with METs higher than 6 include singles

tennis, cycling more than 10 mph, walking more than 4 mph, and cross-country skiing.

The MET system may not be as accurate for overweight or obese adults. Research has shown that using the MET system underestimates the energy used for an activity in this population group. Overweight or obese individuals may end up working at a level too high for them. The therapist is advised to use the rate of perceived exertion (RPE; see Table 12-13) instead.¹¹⁰

As activity level increases, the therapist must monitor heart rate, blood pressure, and fatigue, adjusting activity level accordingly. During phase I (acute hospital) care, the heart rate should not rise more than 25% above resting level, and blood pressure must not rise more than 25 mm Hg above resting level.

When systolic blood pressure falls or fails to increase as the intensity of exercise increases, exercise intensity should be immediately reduced. A drop in systolic blood pressure during exercise below the rest value as measured in the standing position is associated with increased risk of lethal arrhythmia in clients with a prior MI or myocardial ischemia.

Supplemental oxygen may be used to supply the myocardium with oxygen when the demand exceeds supply, thereby reducing myocardial stress and eliminating dyspnea. Blood gas analysis is usually performed within 1 hour of initiating oxygen therapy to establish a baseline of arterial saturation. By monitoring blood gases, oxygen dose can be altered to regulate blood gases and acid-base balance. The therapist must monitor oxygen saturation levels during exercise or intervention, because these activities may increase myocardial oxygen demand (see Table 40-18).

The client with chronic obstructive pulmonary disease (COPD) who receives oxygen therapy must be monitored very closely for symptoms of decreased ventilation, such as headache, giddiness, tinnitus (ringing in the ears), nausea, weakness, and vomiting. Frequently persons with COPD retain carbon dioxide (CO_2), making the use of oxygen deadly. CO_2 levels are elevated in the COPD client, eliminating the drive to breathe normally that is initiated by rising levels of CO_2 . The only drive to breathe in the COPD client is hypoxia (reduced oxygen levels). The administration of oxygen to a person with CO_2 retention can further depress the respiratory drive, resulting in death.

Exercise after Myocardial Infarction

As little as 15 years ago, exercise was avoided following a heart attack, but research has shown that a reasonable amount of regular exercise is the best way to strengthen the heart and control blood pressure, cholesterol, diabetes, and weight. Survivors who exercise usually require less medication, are less likely to need future invasive procedures (e.g., angioplasty, bypass graft), and are less likely to die of a second heart attack than those who remain sedentary.

Traditionally, isometric exercises have been contraindicated, and resistance training or weightlifting has been excluded from the cardiac client's program. Although weight training is not an isometric (static) exercise, it is similar during maximal lifts.

A static muscle contraction that involves 70% or more of maximal effort results in a disproportionate increase in heart rate and blood pressure for the absolute level of oxygen uptake, which is potentially harmful for the ischemic heart.¹⁷⁶ For some people, use of a cane or walker is an isometric use of muscles that can increase heart rate; therefore, careful monitoring of vital signs and indications of perceived exertion is required.

Now, low-risk cardiac clients have undertaken supervised and prescribed weight-training programs without ill effects, especially if regimens incorporate moderate levels of resistance and high numbers of repetitions.^{163,315} Thrombolytic agents reduce the client's blood-clotting ability, necessitating special care to avoid tissue trauma during therapy (e.g., resistive exercises, soft tissue or scar mobilization).

Heart attack survivors are often people who have never exercised before and need sound advice and careful supervision by a physical therapist. Exercise may induce cardiac arrhythmias during diuretic and digitalis therapy, and recent ingestion of caffeine may exacerbate arrhythmias. Exercise-induced arrhythmias are generated by enhanced sympathetic tone, increased myocardial oxygen demand, or both. The immediate postexercise period is particularly dangerous because of high catecholamine levels associated with a generalized vasodilation. Sudden termination of muscular activity is accompanied by diminished venous return and may lead to a reduction in coronary perfusion while heart rate is elevated. A careful cool-down period is required with continued monitoring of vital signs after exercise.

Sexual Activity

People with cardiac disease, both men and women, are prone to sexual dysfunction. Often their concerns are voiced to the therapist. The link between cardiovascular disease and erectile dysfunction in men has been the subject of recent studies. Erectile dysfunction is an early predictor of CAD and should be medically evaluated.^{286,332}

Problems may be caused by medications, anxiety, depression, or limited physical capacity. Hypertensive medications are the most common drugs to cause sexual dysfunction (e.g., loss of sexual desire or ability to reach orgasm).

Marijuana increases myocardial oxygen consumption and heart rate and results in decreased testosterone levels and decreased libido. Cocaine can hinder erection, ejaculation, and orgasm; it may also cause coronary artery vasoconstriction and resulting chest pain and fatal MI.

Fear of death during sexual intercourse, fear of another infarction caused by sexual activity, and diminished sexual ability caused by illness and aging may occur. The sexual partner may have many similar fears and may want to be included in any information provided about return to sexual function. The relative risk of triggering an MI by sexual activity is less than 1%.²³²

Sexual intercourse with orgasm is physiologically equivalent to activities such as a brisk walk or climbing

a flight of stairs. It has been equated to 5 METs of work on an exercise stress test; preorgasmic and postorgasmic phases require about 3.7 METs. Advice to clients should be based on consultation with the physician.

Some general guidelines include the following: (1) when the client can sustain a heart rate of 110 to 120 beats/min with no shortness of breath or anginal pain, he or she can resume sexual activity; (2) sexual activity should be resumed gradually and only after activities such as walking moderate distances (equivalent to 3 or 4 miles on a level treadmill) or climbing stairs comfortably have been accomplished; (3) sexual activity causes the least amount of stress when it occurs in familiar surroundings with the usual partner in a comfortable environment; (4) gradual foreplay helps the heart prepare for coitus; less strenuous sexual activities, such as cuddling, kissing, touching, and hugging, can be engaged in without sexual intercourse; (5) positions requiring isometric contractions should be avoided; (6) eating a large meal or drinking alcohol 1 to 3 hours before sexual activity should be avoided; (7) anal stimulation and anal intercourse should be avoided, because this stimulates the vagus nerve and may cause chest pain and slows down the heart rate and rhythm, impulse conduction, and coronary blood flow²¹⁵; and (8) the physician should be asked about whether the client should take prophylactic nitroglycerin before intercourse.¹⁸⁰

Congestive Heart Failure

Definition and Overview

CHF is a condition in which the heart is unable to pump sufficient blood to supply the body's needs. Backup of blood into the pulmonary veins and high pressure in the pulmonary capillaries lead to subsequent pulmonary congestion and pulmonary hypertension. Failure may occur on both sides of the heart or may predominantly affect the right or left side. Heart failure is not a disease but rather represents a group of clinical manifestations caused by inadequate pump performance from either the cardiac valves or the myocardium. It may be chronic over many years, requiring management by oral medications, or it may be acute and life-threatening, requiring more dramatic medical management to maintain an adequate cardiac output.

Four distinct types of CHF have been recognized: (1) systolic heart failure (caused by contractile failure of the myocardium), (2) diastolic failure (occurs when increased filling pressures are required to maintain adequate cardiac output despite normal contractile function), (3) left-sided heart failure (occurs when the left ventricle can no longer maintain a normal cardiac output), and (4) right-sided heart failure (right-sided ventricular dysfunction secondary to either left-sided heart failure or to pulmonary disease).

Strictly classified, left ventricular failure is referred to as CHF; acute right ventricular failure, seen almost exclu-

Table 12-11 Etiologic and Risk Factors Associated with Congestive Heart Failure

Etiologic Factors	Risk Factors*
Hypertension	Emotional stress
Coronary artery disease	Physical inactivity
Myocardial infarction	Obesity
Valvular heart disease	Diabetes mellitus
Congenital heart disease	Nutritional deficiency (vitamin C, thiamin)
Endocarditis	Fever
Pericarditis	Infection
Myocarditis	Anemia
Cardiomyopathy	Thyroid disorders
Chronic alcoholism	Pregnancy
Atrioventricular malformation	Paget's disease
Thyrotoxicosis (arrhythmia)	Pulmonary disease
Chronic anemia	Medications (e.g., steroids, NSAIDs)
	Drug toxicity
	Renal disease

NSAIDs, Nonsteroidal antiinflammatory drugs.

*Risk factors for new onset or exacerbation of previous congestive heart failure.

sively in association with massive pulmonary embolism, is labeled cor pulmonale. Cor pulmonale is heart disease, but it arises from an underlying pulmonary pathologic condition; therefore it is discussed in Chapter 15. Right-sided heart dysfunction secondary to left-sided heart failure, vascular dysfunction, or congenital heart disease is excluded in the definition of cor pulmonale (see the section on Cor Pulmonale in Chapter 15).

Incidence

CHF is a common complication of ischemic and hypertensive heart disease, occurring most often in the older adult and, in its chronic form, referred to as a cardiogeriatric syndrome. Because the heart muscle is damaged during a heart attack, many heart attack survivors develop CHF. In the United States, heart failure develops in an estimated 500,000 individuals annually: it is the most common cause for hospitalization in people older than 65 years, with an estimated 5 million men and women living with CHF in the United States today. This condition is on the increase as the population ages and more people survive heart attacks.

Etiologic and Risk Factors

Many cardiac conditions predispose individuals to CHF, but hypertension is one of the most prevalent (Table 12-11). People with preexisting heart disease are at greatest risk for the development of CHF, because when the heart is stressed, compensatory mechanisms may be inadequate. For example, a faster redistribution of blood volume and increased demand for oxygen by the myocardium occur with increased activity, such as exercise, resulting in heart failure.

Pulse pressure appears to be the best single measure of blood pressure for predicting mortality in older people and helps explain apparently discrepant results for low

diastolic blood pressure. Pulse pressure is more predictive than even systolic blood pressure alone. Each elevation of 10 mm Hg between systolic and diastolic blood pressure increases the risk of CHF by 14%.^{70,127,252} Although the literature supports the use of pulse pressure as a significant prognostic indicator, day-to-day clinical use is not common.

CHF occurring during middle age as distinguished from CHF at advanced age includes an increasing proportion of women, a shift from CHD to hypertension as the most common etiology, and intact left ventricular systolic function.²⁷⁶ Women tend to have more risk factors and concurrent medical problems, such as hypertension, diabetes, or renal insufficiency. In addition, there may be other gender differences contributing to the development of CHF in women, such as differences in myocardial distensibility (the degree to which muscle fibers stretch) or hormonal differences as yet undetermined.

Paget's disease causes vascular proliferation in the bones. When the disease involves over one third of the skeleton, a high cardiac output state exists and may tax the compromised heart. Medications such as steroids or NSAIDs and drug toxicity are also risk factors. For the person with chronic, stable heart failure, acute exacerbations may occur caused by alterations in therapy, client noncompliance with therapy, excessive salt and fluid intake, arrhythmias, excessive activity, PEs, infection, or progression of the underlying disease.

Pathogenesis and Clinical Manifestations

Over the last 15 years, major advances have occurred in our understanding of heart failure, involving the complex interactions that take place among the adrenergic nervous system, the renin-angiotensin axis, the immune system, the peripheral circulation, and other vasoactive substances in response to impaired cardiac function.

The pathophysiology involves structural changes such as loss of myofilaments, apoptosis (programmed cell death), disturbances in calcium homeostasis, and alteration in receptor density, signal transduction, and collagen synthesis. A neurohormonal hypothesis has replaced the hemodynamic model focusing on the neuroendocrine activation of a progressive disorder of left ventricular remodeling. This cascade of events occurs as a result of a cardiac event (e.g., MI) that develops into a clinical syndrome characterized by impaired cardiac function and circulatory congestion.¹¹³

CHF is a complex event involving one or both ventricles. This discussion is based on left ventricular failure. See the section on Cor Pulmonale in Chapter 15 for a complete discussion of right-sided heart failure. When the heart fails to propel blood forward normally (such as occurs with left ventricular failure), the body utilizes three neurohormonal compensatory mechanisms; these are effective for a short time but eventually become insufficient to meet the oxygen needs of the body.

First, the failing heart attempts to maintain a normal output of blood by enlarging its pumping chambers so that they can hold a greater volume of blood. This lengthening of the muscle fibers, called *ventricular dilation*, increases the amount of blood ejected from the heart. This compensatory mechanism has limits, because con-

tractility of ventricular muscle fibers ceases to increase when they are stretched beyond a certain point.

During this *first compensatory phase*, the right ventricle continues to pump more blood into the lungs. Congestion occurs in the pulmonary circulation with accumulation of blood in the lungs. The immediate result is shortness of breath (most common symptom), and if the process continues, actual flooding of the air spaces of the lungs occurs, with fluid seeping from the distended blood vessels; this is called *pulmonary congestion* or *pulmonary edema*. Congestion in the vascular system interferes with the movement of body fluids in and out of the various fluid compartments, resulting in fluid accumulation in the tissue spaces and progressive edema.

During the *second compensatory phase*, the sympathetic nervous system responds to increase the stimulation of the heart muscle, causing it to pump more often. In response to failing contractility of the myocardial cells, the sympathetic nervous system activates adaptive processes that increase the heart rate and increase its muscle mass to strengthen the force of its contractions. This results in ventricular hypertrophy and a need for more oxygen.

Eventually, the coronary arteries cannot meet the oxygen demands of the enlarged myocardium, and the person may experience angina pectoris owing to ischemia. Secondary compensatory mechanisms activate the sympathetic nervous system and release endothelin from vascular linings, vasopressin (antidiuretic hormone [ADH]) from the pituitary gland, and atrial natriuretic hormone from the heart.

The *third compensatory phase* involves activation of the renin-angiotensin-aldosterone system. With less blood coming from the heart, less blood passes through the kidneys. The kidneys respond by retaining water and sodium in an effort to increase blood volume, which further exacerbates tissue edema. The expanded blood volume increases the load on an already compromised heart. These mechanisms are responsible for the symptoms of diaphoresis, cool skin, tachycardia, cardiac arrhythmias, and oliguria (reduced urine excretion).

When the combined efforts of these three compensatory mechanisms achieve a normal level of cardiac output, the client is said to have compensated CHF. Ultimately, however, the body's efforts to compensate may backfire and produce higher blood volume, higher blood pressure, and more stress on the already weakened heart. The heart's ongoing failure to supply the body with blood compels the body to keep compensating in ways that further burden the heart, and the cycle perpetuates itself. When these mechanisms are no longer effective and the disease progresses to the final stage of impaired heart function, the client has decompensated CHF.

Decompensated CHF ranges from mild congestion with few symptoms to life-threatening fluid overload and total heart failure (Table 12-12). Symptoms usually develop very gradually so that many people do not recognize or report signals of serious disease. The older adult in particular may wrongly associate early symptoms with a lack of fitness or consider them a sign of aging. Confusion and impaired thinking can characterize heart failure in older adults.

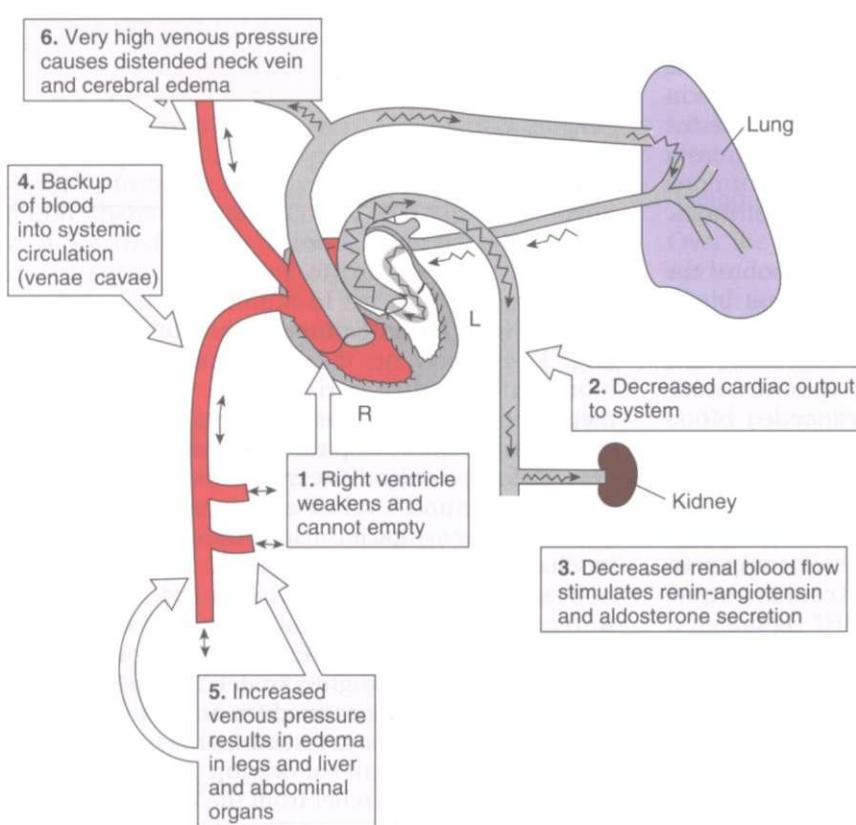
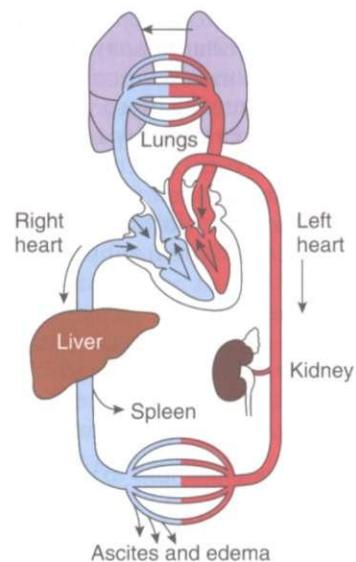
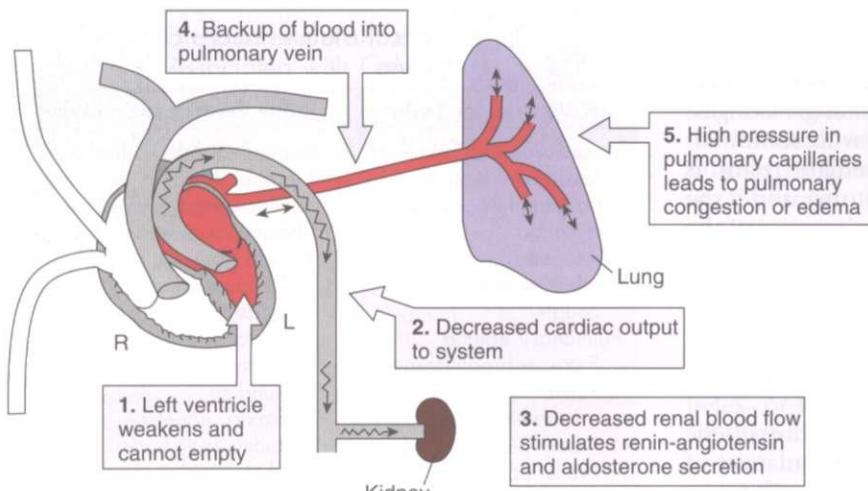
Table 12-12 Clinical Manifestations of Heart Failure

Left Ventricular Failure	Right Ventricular Failure
Progressive dyspnea (exertional first)	Dependent edema (ankle or pretibial first)
Paroxysmal nocturnal dyspnea	Jugular venous distention
Orthopnea	Abdominal pain and distention
Productive spasmodic cough	Weight gain
Pulmonary edema	Right upper quadrant pain (liver congestion)
Extreme breathlessness	Cardiac cirrhosis
Anxiety (associated with breathlessness)	Ascites
Frothy pink sputum	Jaundice
Nasal flaring	Anorexia, nausea
Accessory muscle use	Cyanosis (nail beds)
Rales	Psychologic disturbances
Tachypnea	
Diaphoresis	
Cerebral hypoxia	
Irritability	
Restlessness	
Confusion	
Impaired memory	
Sleep disturbances	
Fatigue, exercise intolerance	
Muscular weakness	
Renal changes	

Left-Sided Heart Failure. Failure of the left ventricle (Fig. 12-12) prevents the heart from pumping enough blood through the arterial system to meet the body's metabolic needs and causes either pulmonary edema or a disturbance in the respiratory control mechanisms. The degree of respiratory distress varies with the client's position, activity, and level of emotional or physical stress, but any of the symptoms listed under Pulmonary Edema in Chapter 15 may occur.

Dyspnea is subjective and does not always correlate with the extent of heart failure; exertional dyspnea occurs in all clients to some degree. Time for dyspnea to subside is an indication of progress or deterioration in a client's status, and it can be measured for documentation. Paroxysmal nocturnal dyspnea resembles the frightening sensation of awakening with suffocation. Once the client is in the upright position, relief from the attack may not occur for 30 minutes or longer. The client often assumes a three-point position, sitting up with both hands on the knees and leaning forward. In severe heart failure, the client may resort to sleeping upright in a chair or recliner. Other sleep disturbances may occur from central sleep apnea present in approximately 40% of all adults with heart failure.

Fatigue and **muscular weakness** are often associated with left ventricular failure, since dyspnea develops along with weight gain and a faster resting heart rate, which decrease the person's ability to exercise. Inadequate cardiac output leads to decreased peripheral blood flow and blood flow



B Right-sided congestive heart failure

Figure 12-12

Pathophysiologic mechanisms of congestive heart failure. **A**, Left-sided heart failure leads to pulmonary edema (see text description). **B**, Right ventricular failure causes peripheral edema that is most prominent in the lower extremities. *Inset*, Integration of the pulmonary and systemic circulation. When the heart contracts normally, it pumps blood simultaneously into both loops, but pump failure causes circulatory or pulmonary problems, depending on the underlying pathologic mechanism. (A and B from Gould B: *Pathophysiology for the health professions*, ed 2, Philadelphia, 2002, Saunders, p 286; inset from Damjanov I: *Pathology for the health-related professions*, ed 3, Philadelphia, 2006, Saunders.)

to skeletal muscle. The resultant tissue hypoxia and slowed removal of metabolic wastes cause the person to tire easily. Disturbances in sleep and rest patterns may aggravate fatigue; muscle atrophy is common in advanced CHF.

Renal changes can occur in both right- and left-sided heart failure, but they are more evident with left-sided failure. During the day, the client is upright, decreased cardiac output reduces blood flow to the kidneys, and the formation of urine is reduced (oliguria). Sodium and water not excreted in the urine are retained in the vascular system, adding to the blood volume.

Diminished blood supply to the renal system causes the kidney to secrete renin, stimulating production of angiotensin, which causes vasoconstriction, thereby causing an increase in peripheral vascular resistance, increasing blood pressure and cardiac work, and resulting in worse heart failure. Renin secretion also indirectly stimulates the secretion of aldosterone from the adrenal gland. Aldosterone acts on the renal tubules, causing them to increase reabsorption of sodium and water, further increasing fluid volume. At night, urine formation increases with the recumbent position as blood flow to the kidney improves. *Nocturia* may interfere with effective sleep patterns, which contributes to fatigue as mentioned.

Right-Sided Heart Failure. Failure of the right ventricle (see Fig. 12-12) to adequately pump blood to the lungs results in peripheral edema and venous congestion of the organs. Symptoms result from congestion in the heart's right side and throughout the venous system (see Table 12-12) (see also the section on Cor Pulmonale in Chapter 15).

Dependent edema is one of the early signs of right ventricular failure, although significant CHF can be present in the absence of peripheral edema. In CHF, fluid is retained because the baroreceptors of the body sense a decreased volume of blood as a result of the heart's inability to pump an adequate amount of blood. The receptors subsequently relay a message to the kidneys to retain fluid so that a greater volume of blood can be ejected from the heart to the peripheral tissues. Unfortunately this compounds the problem and makes the heart work even harder, which further decreases its pumping ability, causing a sense of weakness and fatigue.

The retained fluid commonly accumulates in the extra-cellular spaces of the periphery. The resultant edema is usually symmetric and occurs in the dependent parts of the body, where venous pressure is the highest. In ambulatory persons, edema begins in the feet and ankles and ascends up the lower legs (pretibial areas). It is most noticeable at the end of a day and often decreases after a night's rest. In the recumbent person, pitting edema may develop in the presacral area and, as it worsens, progress to the medial thighs and genital area.

Jugular venous distention also results from fluid overload. The jugular veins empty unoxygenated blood directly into the superior vena cava. Since no cardiac valve exists to separate the superior vena cava from the right atrium, the jugular veins give information about activity on the right side of the heart. As fluid is retained



Figure 12-13

Jugular venous distention occurs bilaterally if there is a cardiac cause such as congestive heart failure; a unilateral distention indicates a localized problem. (From Daily EK, Schroeder JP: Techniques in bedside hemodynamic monitoring, ed 2, St Louis, 1981, Mosby.)

and the heart's ability to pump is further compromised, the retained fluid backs up into both the lungs and the venous system, and the jugular veins reveal this. Jugular venous pulsations are examined by inspecting the silhouette of the neck with the person reclining at a 45-degree angle (Fig. 12-13). The right internal jugular vein is recommended because the left internal jugular may be falsely elevated in some people.

As the liver becomes congested with venous blood it becomes enlarged, and *abdominal pain* occurs. If this occurs rapidly, stretching of the capsule surrounding the liver causes severe discomfort, and the person may notice either a constant aching or a sharp *right upper quadrant pain*. In chronic CHF, longstanding congestion of the liver with venous blood and anoxia can lead to ascites (see Fig. 17-5) and jaundice, which are symptoms of liver damage. Anorexia, nausea, and bloating develop secondary to venous congestion of the GI tract. Anorexia and nausea may also result from digitalis toxicity, which is a common problem since digitalis is usually prescribed for CHF.

Cyanosis of the nail beds appears as venous congestion reduces peripheral blood flow. Clients with CHF often feel anxious, frightened, and depressed. Fears may be expressed as frightening nightmares, insomnia, acute anxiety states, depression, or withdrawal from reality.

MEDICAL MANAGEMENT

DIAGNOSIS. Diagnosis is based on the clinical picture and depends on where symptoms are on the continuum of mild to severe. Because the two sides of the heart serve different functions, distinguishing the symptoms of left-sided heart failure from those of right-sided heart failure is critical in both diagnosis and treatment. Equally important is consideration of systolic and diastolic dysfunction, both of which indicate a functional or structural defect in the ventricles.

An echocardiogram is the main diagnostic tool; noninvasive cardiac tests such as ECG and chest radiography are secondary tools that can determine left ventricular size and function well enough to confirm the diagnosis. Cardiac catheterization is not routinely performed, but it may be useful in certain cases (e.g., atherosclerotic heart disease, which is potentially correctable). Arterial blood gases are measured to evaluate oxygen saturation. Liver enzymes (e.g., aspartate transaminase [AST], alkaline phosphatase) are often elevated (see Tables 40-5 and 40-18); liver involvement with hyperbilirubinemia commonly occurs, resulting in jaundice.

A new screening tool for individuals with suspected left ventricular dysfunction has been introduced. Measuring B-type natriuretic peptide, a protein secreted from the cardiac ventricles in response to wall tension and pressure overload, can reliably predict the presence or absence of heart failure, even helping to identify when dyspnea is associated with heart failure or some other underlying pathologic condition.²⁰⁸

PREVENTION AND TREATMENT. Managing heart failure begins with treatment of the underlying cause whenever possible. Nonpharmacologic interventions such as diet and exercise that alter interactions between the heart and the periphery are now accepted therapeutic approaches.

Alterations in lifestyle reduce symptoms and the need for additional medication. There is an urgent need to develop more effective strategies for the prevention and treatment of this increasingly common disorder. Multiple comorbidities in older clients require a multidisciplinary approach to management. Persons with CHF are placed on a sodium-restricted diet, sometimes with limited fluid intake. Emotional and physical rest during the initial phases of intervention is also important in diminishing the workload of the heart.

Activity and Exercise. Traditionally, the diagnosis of CHF was a contraindication for participation in exercise training because of concerns that further decline in cardiac function would occur. It is now clear that activity restriction is no longer appropriate, since exercise programs have proved to quantitatively achieve results similar to those attained with most effective drug therapies. These findings have shifted attention away from treating the heart toward exercising the muscles.

Whenever possible, physical activity and exercise are prescribed per client tolerance. Physical training for clients with CHF results in an increase in muscular strength and better adaptation to effort owing to the effect of training on skeletal muscles (e.g., decreased vascular resistance in the muscles, delay in the onset of anaerobic metabolism). Exercise training has also been shown to improve exercise capacity, reduce symptoms, improve psychosocial status, and improve functional capacity.^{159,266} Recently, resistance training combined with short or long bouts of aerobic exercise was found beneficial for patients with CHF.⁶⁰

Pharmacotherapy. Pharmacologic therapy is now responsive to the updated understanding of CHF as a cascade of neurohormonal events centered on ventricular remodeling. Pharmacologic agents are used to reduce the

heart's workload, increase muscle strength and contraction, and inhibit neuroendocrine responses to heart failure (see Tables 12-5 and 12-6).

ACE inhibitors have become standard therapy for heart failure because of their ability to block the renin-angiotensin-aldosterone system, increasing renal blood flow and decreasing renal vascular resistance, thereby enhancing diuresis. ACE inhibitors reduce left ventricular filling pressure and moderately increase cardiac output. Vasodilator therapy in combination with ACE inhibitors prolongs life in persons with moderate to severe heart failure.

Diuretics are used to control fluid buildup and prevent congestion, and digoxin may be added to stimulate the heart's pumping action if symptoms persist despite treatment with ACE inhibitors and diuretics. Angiotensin II receptor antagonists have been added to function as an antihypertensive and enhance the clearance of sodium and water.

The β -blockers, once rarely considered in the treatment of CHF, have been shown effective in reducing symptoms, improving clinical status, reducing hospitalizations, and reducing the risk of death. Combining β -blockers with ACE inhibitors can produce additive effects on two neurohormonal systems (renin-angiotensin system and sympathetic nervous system).

A new drug, nesiritide (human recombinant B-type natriuretic peptide), has been introduced as a first-line medication for decompensated CHF that inhibits sympathetic activity and dilates arterial and venous vessels. Nesiritide binds to receptors in the vasculature, kidney, and other organs to mimic the vasodilatory and diuretic actions of endogenous natriuretic peptides.⁸²

Surgery. Surgical intervention may include CABG (see Fig. 12-4) for underlying myocardial ischemia and infarction; reconstruction of incompetent heart valves; ventricular remodeling or heart reduction (e.g., Batista procedure, in which a piece of the heart tissue is removed and the heart muscle is sutured back together, making a smaller, tauter heart with a stronger heartbeat); internal counterpulsation (Fig. 12-14) or external counterpulsation, which uses an external pump or balloon to adjust the aortic blood pressure; temporary ventricular assistive devices for people unable to come off bypass (see Chapter 21); and use of an artificial heart or cardiac transplantation.

The implantation of skeletal muscle (removed from the individual's thigh and multiplied in the laboratory) injected into the postinfarction scar after infarction in the case of severe ischemic heart failure has been shown experimentally to improve heart function.²¹⁹ A review of surgical innovations for chronic heart failure in the context of cardiopulmonary rehabilitation for the therapist is available.¹⁵⁸ See also the sections on Atherosclerosis: Treatment in this chapter and Heart Transplantation in Chapter 21.

Cardiac transplantation is now more common for treatment of heart failure. Transplantation is successful for selected individuals, usually those who are treated early in the course of heart failure, before advanced symptoms develop. Reform of the selection process is recommended to identify people who, although not critically

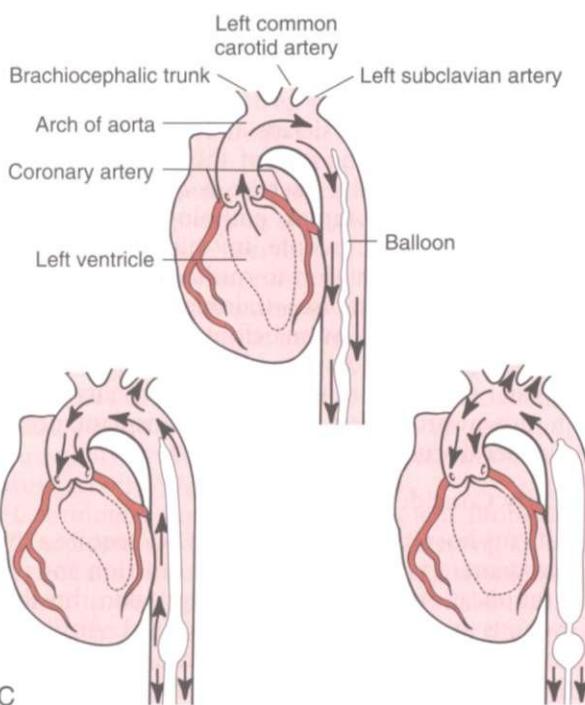
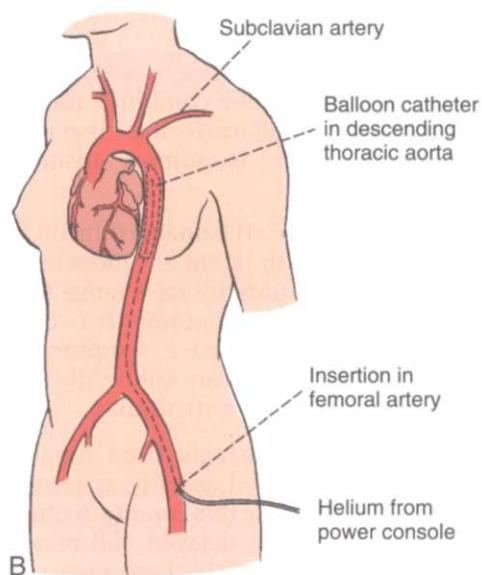


Figure 12-14

The intraaortic balloon pump (IABP) is a common type of cardiac assist device that is used to improve myocardial oxygen supply-demand for individuals with deteriorating hemodynamics or ongoing ischemia, as evidenced by rest pain or electrocardiographic changes in the region of the infarct. The primary functions of balloon counterpulsation are to reperfuse the coronary arteries at the end of systole and reduce the left ventricular afterload (the amount of work the ventricle must do), thereby decreasing myocardial oxygen consumption and improving cardiac output. These intravascular catheter-mounted counterpulsation devices are traditionally used for cases of cardiogenic shock following cardiac surgery or an acute myocardial infarction as well as for people who have chronic end-stage heart failure and who are not candidates for long-term ventricular assistive device (VAD) support. The rationale for IABP counterpulsation in this latter situation is to maintain systemic perfusion and preserve end-organ function until cardiac transplantation occurs. **A**, The catheter is usually placed through the femoral artery, and the balloon is moved up the iliac artery to the descending aorta, where it is then placed. **B**, above the renal arteries and below the subclavian artery. This position is critical in order to prevent ischemia to the upper extremities or kidneys. **C**, When the heart contracts (systole), the balloon is deflated, creating a decline in aortic pressure. After the heart contracts (during diastole), the balloon is filled with air, causing the blood to regurgitate back toward the root of the aorta, thereby perfusing the coronary arteries. When the left ventricle is ready to pump, the balloon is deflated (cardiac systole again), reducing ventricular afterload. [A, courtesy Chris Wells, PT, MS, PhD, University of Pittsburgh Medical Center, 2001. B, from Black JM, Hawks JH, Keene AM: *Medical-surgical nursing: clinical management for positive outcomes*, ed 7, Philadelphia, 2005, Saunders. C, from Lewis SL, Heitkemper MM, Dirksen SR: *Medical-surgical nursing: assessment and management of surgical problems*, ed 7, St Louis, 2007, Mosby.]

ill, will not survive without early transplantation. See further discussion in Chapter 21.

A pacemaker-like device designed to deliver electrical stimulation to the ventricles (biventricular pacing) in an effort to improve the heart's overall cardiac efficiency by coordinating the heart's contractions (both ventricles

pump at the same time, making the heart pump more forcefully) has been approved by the U.S. Food and Drug Administration (FDA). This technique, referred to as cardiac resynchronization therapy, is available on a limited basis for selected individuals with severe heart failure. The results are promising for people who because

of age criteria or lack of donor hearts are not able to undergo cardiac transplantation.

Other similar devices are under continued investigation and development, as is the combined use of resynchronization therapy with pharmacologic therapy and/or a cardioverter-defibrillator as adjunct treatment for CHF.

PROGNOSIS. Treatment of CHF remains difficult, and the prognosis is poor, even with recent advances in pharmacologic therapy. Annual mortality rates range from 10% in stable clients with mild symptoms to over 50% in people with advanced, progressive symptoms. About 40% to 50% of clients with heart failure die suddenly, probably owing to ventricular arrhythmias.

To achieve the maximal benefit from drug therapy, symptoms must be recognized as early as possible and intervention initiated. Because this condition often develops gradually, intervention is delayed, full resolution is not usually possible, and CHF becomes a chronic disorder.

Exercise capacity was the most powerful predictor of survival in CHF, but a new test that measures swings in heart rate during the day has been developed that can identify individuals who are at the highest risk of dying from CHF. The test measures the amount by which the heart rate changes from slow rates to fast rates in one 24-hour period. The less the heart rate varies over 24 hours, the more likely a person is to die of CHF.²⁴⁴

Other signs of poor prognosis include severe left ventricular dysfunction, severe symptoms and limitation of exercise capacity, secondary renal insufficiency, and elevated plasma catecholamine levels.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-7

Congestive Heart Failure

PREFERRED PRACTICE PATTERNS

6A: Primary Prevention/Risk Reduction for Cardiovascular/Pulmonary Disorders

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C, 6F may apply for complications associated with mechanical vascular devices or implants (see Chapter 21).

7A: Primary Prevention/Risk Reduction for Integumentary Disorders (prevent complications of bed rest)

Therapists have a unique role in the prevention, medical management, and rehabilitation of people with heart failure. Physical therapists can provide programs that profoundly improve the exercise tolerance and functional status of individuals with CHF.

Medical intervention can be more objectively implemented by using information obtained during physical therapy assessments and interventions. Tests such as the 6-minute walk test may be helpful in predicting peak oxygen consumption and early survival as well as implementing a proper exercise conditioning program for people with advanced heart failure.⁵⁹

Box 12-10

NEW YORK HEART ASSOCIATION'S FUNCTIONAL CLASSIFICATION OF HEART DISEASE

- **Class I:** Cardiac disease present but no limitation on physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain.
- **Class II:** Slight limitation on physical activity. Comfortable at rest, but ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain.
- **Class III:** Marked limitation of physical activity. Comfortable at rest, but less than ordinary physical activity causes fatigue, palpitation, dyspnea, or anginal pain.
- **Class IV:** Unable to carry on any physical activity without discomfort. Symptoms of cardiac insufficiency or of the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort is increased.

Education of physicians and other health care professionals about the role of the physical therapist in defining prescriptive exercise is important. Consideration for the complex pathologic conditions and comorbidities of people in this population is an important contribution to cardiac rehabilitation from the physical therapist's training. Clients should be referred to rehabilitation before the $V_{O_{2 \text{ max}}}$ drops below 14 ml/kg/min and when the wedge pressure is still greater than 16 mm Hg (i.e., before clients progress in a downward spiral requiring transplantation). See further discussion in Chapter 21.

Early Considerations

Clients hospitalized with severe CHF require a therapy program to maintain pulmonary function and prevent complications of bed rest (e.g., skin breakdown, venous stasis, venous thrombus, PEs). An important aspect of intervention is functional assessment (Box 12-10) and physical exercise within the limitations set by the physician. See also established guidelines for exercise training in CHF.^{58,59} Physical therapy assessment of cardiopulmonary status is beyond the scope of this text. The clinician is referred to any of the specific examination and assessment texts available.^{114,150,162}

The therapist should be aware of psychosocial considerations in older adults with CHF. Neuropsychiatric conditions such as Alzheimer's dementia and complications such as delirium are common in older adults with CHF. Persistent alcohol abuse and cigarette smoking often contribute to the onset and progression of heart failure. Major depression, other depressive disorders, anxiety, and social isolation are common and have adverse effects on functional status, quality of life, and prognosis. Working as a team with psychologists and social workers can address these issues effectively.

Monitoring Vital Signs

Aerobic capacity is likely impaired and even more so if the client is deconditioned; adaptive responses to activity may be attenuated or inadequate; activity may

exacerbate cardiovascular pump dysfunction; and signs of fatigue and shortness of breath are common. The downward cycle of disease, deconditioning, decreased activity, and disability necessitates the monitoring of vital signs.⁵² Progressing activities from bed rest to transfers or ambulation requires vital sign assessment immediately after the major activity and 3 minutes later to assess for return to baseline. Oxygen may be administered by mask or cannula; team members should consult respiratory therapy staff to determine appropriate oxygen levels during exercise.

Monitor the client for signs of increasing peripheral edema by assessing jugular neck vein distention, peripheral edema in the legs or sacrum, and any report of right upper quadrant pain. In the outpatient or home health setting, the client is advised to call the nurse or physician if shoes, belt, or pants become too tight to fasten, usual activities of daily living or tasks become difficult, extra sleep is needed, or urination at night becomes more frequent.

Monitoring blood pressure is essential to detect heart failure; observe for decreasing blood pressure and report any change in status to the nurse or physician immediately. Observe for flat or falling systolic blood pressure in response to activity indicative of inadequate pump function (a linear increase of systolic blood pressure with increased activity should be seen). Exaggerated increases in heart rate may be observed as the heart attempts to maintain adequate cardiac output. Observe for dyspnea at rest and/or with activity, and auscultate for changes from baseline during activity.⁵² See also Appendix B.

Continuous supervision and frequent monitoring of blood pressure are necessary when starting an exercise program for someone with CHF. RPE should range from 11 to 14 (light to somewhat hard; Table 12-13). Anginal symptoms should not exceed 2 on the 0 to 4 angina scale (moderate to bothersome), and exertional dyspnea should not exceed a rating of mild, some difficulty with activity. Initially, full resuscitation equipment should be available.⁴³

Positioning

Positioning is important, and the client is taught to use a high Fowler's position (head of the bed elevated at least 20 inches above the level) or chair to reduce pulmonary congestion, facilitate diaphragmatic expansion and ventilation, and ease dyspnea.

The legs are maintained in a dependent position as much as possible to decrease venous return. Range of motion to decrease venous pooling and monitoring for the development of thrombophlebitis (e.g., unilateral swelling, calf pain, pallor) are required.

Exercise and Congestive Heart Failure

The American College of Sports Medicine's guidelines¹¹ suggest that CHF clients entering an exercise program should start with moderate-intensity exercise (40% to 60% $\dot{V}O_{2\max}$) for a duration of 2 to 6 minutes, followed by 2 minutes of rest. Blood pressure and heart rhythm should be routinely monitored at rest, during peak exercise, and after cool-down. The goal is to gradually

Table 12-13 Perceived Exertion Scale*

10-Grade Scale	Verbal Rating
0	No exertion at all
0.5	Extremely light
1	Very light
2	Light
3	Moderate
4	Somewhat hard
5	Hard
6	
7	Very hard
8	
9	
10	Very, very hard
—	Maximal exertion

Modified from Borg GA: Psychosocial bases of perceived exertion, *Med Sci Sports Exerc* 472:194-381, 1982.

*Using a perceived exertion scale is a useful approach to activity prescription. The individual is asked to identify a desirable rating of perceived exertion and uses that level of intensity as a daily guideline for activity. A suggested rating of perceived exertion for most healthy individuals is 3 to 5 (moderate to hard on the 10-grade scale); for the compromised person, a more moderate level of perceived exertion may be recommended by the physician.

increase the intensity and duration of exercise. Others advocate starting CHF clients at a low to moderate exercise intensity (less than 40% $\dot{V}O_{2\max}$) with a shorter duration of exercise initially and a shorter rest period of less than 2 minutes). Recommendations for interval exercise training (following work phases of 30 seconds by recovery phases of 60 seconds) have also been reported.²²⁰

The best guideline is to customize initial exercise intensity for each individual,³⁵³ keeping in mind the individual's goals and expected outcomes (e.g., preparation for transplantation, improved functional daily living, perceived quality of life). Exercise should be avoided immediately after eating or after taking vaso-dilator medication. Using an interval training approach is helpful with those individuals who demonstrate marked exercise intolerance.

Symptoms and general fatigue level serve as a guideline to determine frequency, and warm-up/cool-down periods should be longer than normal for observation of possible arrhythmias. Determination of appropriate exercise intensity based on 40% to 60% $\dot{V}O_{2\max}$ is recommended (rather than based on heart rate peak) because the response to exercise is frequently abnormal in people with CHF. Alternately, the initial exercise intensity should be 10 beats below any significant symptoms, including angina, exertional hypotension, arrhythmias, and dyspnea.

Rehabilitation personnel must observe for symptoms of cardiac decompensation during exercise, including cough or dyspnea, hypotension, light-

Continued.

headedness, cyanosis, angina, and arrhythmias. Exercise progression following these recommendations is available in detail⁴³; see also Monitoring Vital Signs in this section. Endurance exercise training has been shown to modify neuroendocrine activation in heart failure and may have a long-term beneficial impact.⁴⁴

The therapist should keep in mind that some older CHF clients are unable to increase their exercise intensity or duration despite starting very slowly. These people do not achieve the goal of increased endurance and often leave the program owing to increased symptoms and exercise intolerance. Maintaining or even improving functional activities and independence at home may be more appropriate goals for this group. An excellent review of exercise assessment, exercise training, and exercise training guidelines in heart failure for all clients is available.^{57,58}

Medications

Diuretics can produce mild to severe electrolyte imbalance requiring special consideration (see Chapter 5). A small drop in the serum potassium level can precipitate digoxin poisoning (digitalis toxicity) and serious arrhythmias. This situation is a life-threatening condition that occurs in one of every five clients and may present with systemic or cardiac manifestations.

Any sign or symptom of digitalis toxicity should be reported to the physician (see Table 12-5). Digitalis toxicity can cause a dip in the ST segment on ECG; whenever possible, the ECG should be monitored during exercise. Activity should not increase the magnitude of the altered ST segment. Side effects from digitalis can occur when digitalis levels are within the therapeutic range (less than 2.0 ng/ml), but the albumin level will be low (less than 3.5 g/dl; see Table 40-5) because digitalis binds to albumin in the serum.

If the serum albumin level is low, digitalis may not be bound to the albumin-binding sites and will be circulating as "free" digoxin. Watch for a low, irregular pulse (less than 60 beats/min); the heart rate normally increases to compensate for CHF, but in the presence of digoxin, heart rate decreases.

NSAIDs, including over-the-counter drugs such as ibuprofen, increase fluid retention independently and significantly blunt the action of diuretics and other cardiovascular drugs (especially ACE inhibitors), exacerbating preexisting CHF and causing isolated lower extremity edema. The major consideration for exercise in clients taking ACE inhibitors is the possibility of hypotension and accompanying arrhythmias. These problems should be reported to the physician and can be addressed by maintaining proper hydration and by altering dosages and the simultaneous use of other medications.

Orthostatic (Postural) Hypotension

Definition and Overview

The term *orthostatic (postural) hypotension* signifies a decrease of 20 mm Hg or greater in systolic blood pressure or a drop of 10 mm Hg or more in both systolic and diastolic arterial blood pressure with a concomitant pulse increase of 15 beats/min or more on standing from a supine or sitting position.

Orthostatic hypotension may be acute and temporary or chronic. Orthostatic hypotension occurs frequently in older adults and occurs in more than one half of all frail, older adults, contributing significantly to morbidity from syncope, falls, vital organ ischemia (e.g., MI, transient ischemic attacks), and mortality among older adults with diabetic hypertension. It is highly variable over time but most prevalent in the morning when supine blood pressure is highest and on first arising.

Etiologic Factors

Orthostatic hypotension is recognized in all groups as a cardinal feature of autonomic nervous dysfunction as well as other nonneurogenic etiologies (Box 12-11). In young adults, orthostatic intolerance and tachycardia may be associated with norepinephrine transporter deficiency. A single gene coding a protein that clears norepinephrine does not function in some individuals, pointing to a genetic etiology.

Postural reflexes are slowed as part of the aging process for some, but not all, persons. Normal aging is associated with various changes that may lead to postural hypotension. Cardiac output falls with age; in the older adult with hypertension, it is even lower. When older subjects (more than 65 years) are put under passive postural stress (60-degree upright tilt), their stroke volume falls even further. These normal changes obviously predispose the aging adult to postural hypotension from any process that further reduces fluid volume or vascular integrity. For example, pooling of blood after eating may lead to profound hypotension, called *postprandial hypotension*.

Box 12-11

CAUSES OF ORTHOSTATIC HYPOTENSION

- Volume depletion (e.g., burns, diabetes mellitus, sodium or potassium depletion)
- Venous pooling (e.g., pregnancy, varicosities of the legs)
- Side effects of medication (e.g., antidepressants, antihypertensives; see Table 12-5)
- Prolonged immobility
- Starvation, malnutrition, eating disorders
- Performing Valsalva maneuver
- Sluggish normal regulatory mechanisms (e.g., anatomic variation, altered body chemistry)
- Autonomic nervous system dysregulation (e.g., diabetes mellitus, Parkinson's disease, aging, fibromyalgia syndrome, chronic renal failure)

In addition, as systolic pressure rises from atherosclerosis, baroreceptor sensitivity and vascular compliance are reduced further, increasing the likelihood of postural hypotension. In the older adult with hypertension and cardiovascular disease receiving vasoactive drugs, the circulatory adjustments to maintain blood pressure are disturbed, leaving the person vulnerable to postural hypotension.¹⁷⁸

Drugs are a major cause of orthostatic hypotension in the aging adult. Many have effects on the autonomic nervous system, both centrally and peripherally, and on fluid balance. Diuretics, calcium channel blockers, nitrates, and L-dopa have hypotensive effects. Antidepressants are a common, overlooked cause of orthostasis, even though this is a known side effect of these medications. A general result of treatment for hypertension may be hypotension. In addition, many older adults with systolic hypertension have postural hypotension that may require management before the hypertension is addressed.

Chronic orthostatic hypotension may occur secondary to a specific disease, such as endocrine disorders, metabolic disorders, nephropathy, or neurogenic disorders affecting the autonomic or central nervous systems. Alcohol and drugs such as vincristine used in the treatment of cancer can cause autonomic neuropathy.

Pathogenesis

Orthostasis is a physiologic stress related to upright posture. When a normal individual stands up, the gravitational changes on the circulation are compensated for by several mechanisms, including the circulatory and autonomic nervous systems. On standing, the force of gravity in the vertical axis causes venous pooling in the lower limbs, a sharp decline in venous return, and reduction in filling pressure of the heart, which increase further on prolonged standing because of shifting of water to interstitial spaces and hemoconcentration.

These mechanical events can cause a marked reduction in cardiac output and consequent fall in arterial blood pressure. In healthy people, cardiac output and blood pressure regulation are maintained by powerful compensatory mechanisms involving a rise in heart rate. Blood pressure is maintained by a rise in peripheral resistance. These compensatory mechanisms are initiated by the baroreceptors located in the aortic arch and carotid bifurcation. Orthostatic hypotension results from failure of the arterial baroreflex, most commonly because of disorders of the autonomic nervous system.¹⁷⁸

In people with autonomic failure or dysreflexia (e.g., Parkinson's disease, aging, diabetes, fibromyalgia), orthostatic hypotension results from an impaired capacity to increase vascular resistance during standing. This dysfunction leads to increased downward pooling of venous blood and a consequent reduction in stroke volume and cardiac output that exaggerates the orthostatic fall in blood pressure.

Approximately 80% of the blood pooled in the lower limb is contained in the upper leg (thighs, buttocks) with less pooling in the calf and foot. The location of the

additional venous pooling has not been clearly identified, but present data suggest the abdominal compartment and perhaps leg skin vasculature. The pooled blood in the veins of the feet and calves is arterial in origin in that it arises as a result of decreased venous drainage of that region.

In contrast, the blood pooled in the thighs, buttocks, pelvis, and abdomen arises primarily from venous reflux. The pooled blood is not actually stagnant; its mean circulatory time through the dependent region is merely increased by changes in the pressure gradient across the vascular bed and by increases in venous volume. The identification of venous pooling may offer insights for intervention techniques in the future.

Clinical Manifestations

Orthostatic hypotension is often accompanied by dizziness, blurring or loss of vision, and syncope or fainting. There are three main modes of presentation in the older adult: (1) falls or mobility problems, (2) acute or chronic mental confusion, and (3) cardiac symptoms.

A common clinical picture is the person whose legs give way when attempting to stand, usually after prolonged recumbency, after physical exertion, or in a warm environment. These episodes may be accompanied by confusion, pallor, tremor, and unsteadiness. Loss of consciousness may cause frequent falls and additional injuries that can be quite serious. Ischemic neck pain in the suboccipital and paracervical region is often reported by individuals with autonomic failure and orthostatic hypotension.⁴¹

Other reported ischemic symptoms of orthostatic hypotension are nonspecific, such as lethargy, weakness, low backache, calf claudication, and angina. Some older adults may experience unexpected and unexplained falls associated with orthostatic hypotension. The cause of such falls may be circulatory impairment that results in a drop in blood pressure on standing upright quickly. Orthostatic hypotension may be an early sign of some other illness or the effects of medication.

Medical Management

There are several general and specific approaches to the management of orthostatic hypotension but no curative intervention for orthostatic hypotension of unknown cause. Prevention is important, and whenever the underlying disorder causing hypotension is corrected, symptoms cease. Nonneurogenic causes, such as diminished intravascular volume, are treated specifically. In orthostatic hypotension caused by autonomic failure there are considerable difficulties in reestablishing sympathetic or parasympathetic efferent activity.

Tilt study or tilt-table testing may be used to assess hypotension by monitoring blood pressure and pulse while tilting a person from horizontal supine to 60 degrees upright. This test has proved very valuable in determining the cause of dizziness or syncope and can reveal irregularities in the vascular regulating system. A combination of general measures and pharmacologic measures is needed in the management of neurogenic postural hypotension.¹³⁴

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-8

Orthostatic Hypotension**PREFERRED PRACTICE PATTERNS**

5A: Primary Prevention /Risk Reduction for Loss of Balance and Falling (side effects of medication)

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

Many medications used to treat hypertension can result in hypotension, especially when combined with interventions or exercise that results in vasodilation. Of particular concern are heat modalities, such as the whirlpool or Hubbard tank. In addition, moderate to vigorous exercise of large muscle groups can produce significant vasodilation and can result in hypotension. This is particularly true following exercise, when venous return diminishes as exercise abruptly ceases. A cool-down period is essential, and safety measures must be employed.³⁴⁶ Aerobic conditioning is an important part of treatment for orthostatic hypotension resulting from autonomic insufficiency, perhaps best accomplished through aquatic exercise therapy.¹³⁴

Stationary standing, as is performed in many activities of daily living, can produce hypotension, especially among those individuals with autonomic failure. With autonomic failure, symptoms of postural hypotension are increased on standing after exercise. The therapist can instruct the individual in protective measures that reduce excessive orthostatic blood pooling, including avoidance of precipitating factors.

Anyone with orthostatic hypotension, especially persons taking antihypertensive agents, should be instructed to rise slowly from the bed or chair after a long period of recumbency or sitting to avoid loss of balance and prevent falls. Dorsiflexing the feet (ankle pumps), raising the arms overhead with diaphragmatic breathing, and abdominal compression before standing often promote venous return to the heart, accelerate the pulse, and increase blood pressure.

The use of abdominal binders and elastic stockings may also help with venous return. Stockings should not be taken off at night to avoid falls when getting up to go to the bathroom or when getting out of bed in the morning. Elevating the head of the bed 5 to 20 degrees prevents the nocturnal diuresis and supine hypertension caused by nocturnal shifts of interstitial fluid from the legs to the rest of the circulation. Eating small meals may help to avoid postprandial (after eating) hypotension.

The person who becomes hypotensive should assume a supine position with legs elevated to increase venous return and to ensure cerebral blood flow. As previously mentioned, this position must be monitored carefully for anyone with orthostatic hypotension and CHF, possibly requiring modifying the position to include slight head and upper body elevation. Crossing the legs, which involves contraction of the agonist and antagonist muscles, also has been shown to increase cardiac output, thereby increasing blood pressure.³¹¹

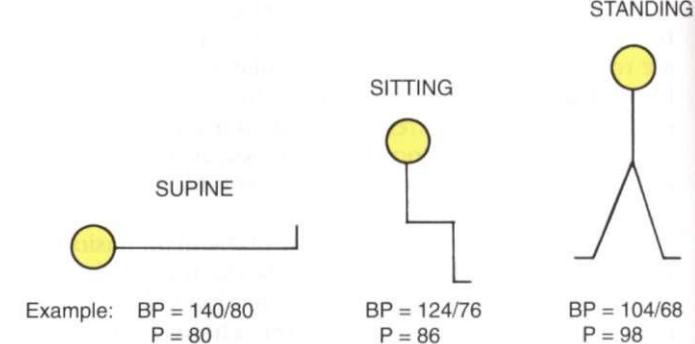


Figure 12-15

Assessing postural hypotension. After measuring the blood pressure (BP) and pulse (P) in the supine position, leave the blood pressure cuff in place and assist the person in sitting. Remeasure the blood pressure within 15 to 30 seconds. Assist the person in standing, and measure again. A drop of more than 20 mm Hg systolic and more than 10 mm diastolic accompanied by a 10% to 20% increase in heart rate (pulse) indicates postural hypotension. Sample measurements are given. (From Black JM, Hawks JH, Keene AM: *Medical-surgical nursing: clinical management for positive outcomes*, ed 7, Philadelphia, 2005, Saunders.)

The physician should be notified if the person remains symptomatic after these measures have been taken. Anyone who is considered borderline hypotensive when tested in the supine position should have blood pressures measured and pulses counted in a sitting position with the legs dangling. If no change occurs when this is done, repeat the measurements with the person standing, if possible. A drop in systolic pressure of 10 to 20 mm Hg or more associated with an increase in pulse rate of more than 15 beats/min suggests depleted intravascular volume (Fig. 12-15). Some normovolemic persons with peripheral neuropathies or those taking antihypertensive medications may demonstrate an orthostatic fall in blood pressure but without an associated increase in pulse rate.

Myocardial Disease**Myocarditis**

Myocarditis is a relatively uncommon acute or chronic inflammatory condition of the muscular walls of the heart (myocardium). It has now been reclassified by the American Heart Association as an acquired (inflammatory) cardiomyopathy.²¹²

It is most often a result of bacterial or viral infection, but it also includes those inflammatory processes related to infectious and noninfectious causes of ischemic heart disease. Other possible causes of myocarditis include chest radiation for treatment of malignancy, sarcoidosis, and drugs, such as lithium, interleukin-2, and cocaine.

The therapist is most likely to treat the person with systemic lupus erythematosus (SLE) (see Chapter 7) who may have a type of myocarditis called *lupus carditis* (see also the section on The Heart in Collagen Vascular Diseases: Lupus Carditis in this chapter). SLE is a multisys-

tern autoimmune disease characterized by a release of autoantibodies into the circulation, with a subsequent inflammatory process that can target the heart and vasculature.

Myocarditis typically evolves through active, healing, and healed stages that are characterized by inflammatory cell infiltrates leading to interstitial edema and focal myocyte necrosis with replacement fibrosis over time. Ventricular tachyarrhythmias develop as a result of the pathologic changes¹ creating an electrically unstable environment.²¹²

Clinical evidence of cardiac involvement is found in up to one half of all people with SLE. Clinical manifestations may include mild continuous chest pain or soreness in the epigastric region or under the sternum, palpitations, fatigue, and dyspnea; and onset may follow a viral upper respiratory tract illness in the population at large as well as in persons with SLE. Complications include heart failure, arrhythmias, dilated (congestive) cardiomyopathy (see next section), and sudden death.

Myocarditis usually resolves with treatment of the underlying condition or cause; specific antimicrobial therapy is prescribed if an infectious agent can be identified. Viral myocarditis is treated with medications that improve cardiac output and reduce arrhythmias, if present. Management of myocarditis in SLE is usually with corticosteroids, but immunosuppressive agents may be required. Myocarditis that progresses to dilated cardiomyopathy with heart failure is frequently fatal without heart transplantation.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-9

Myocarditis

PREFERRED PRACTICE PATTERNS

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

Other patterns may apply depending on underlying disease (e.g., SLE, cancer).

Active myocarditis is considered a contraindication for therapy, because this condition can progress very quickly and stress must be avoided; each case is evaluated by the physician. Athletes in whom myocarditis is suspected or diagnosed should discontinue all sports for 6 months after onset of symptoms. Preparticipation evaluation of cardiac function is essential before resuming sports activities. An athlete can resume competing when ventricular function and cardiac dimensions return to normal and clinically significant arrhythmias are absent on ambulatory monitoring.⁹⁴

If an impairment of myocardial contractility is present, diastolic blood pressure may be elevated to maintain stroke volume. Disruptions leading to lethal cardiac arrhythmias cannot be predicted. See also Special Implications for the Therapist: Infective Endocarditis and Special Implications for the Therapist: Rheumatic Fever and Heart Disease.

Cardiomyopathy

Definition and Overview. Cardiomyopathy is actually part of a group of conditions affecting the heart muscle itself, so that contraction and relaxation of myocardial muscle fibers are impaired. The original definition of *cardiomyopathy* stated that this condition was not caused by other heart or systemic disease, which excluded structural and functional abnormalities caused by valvular disorders, CAD, hypertension, congenital defects, and pulmonary vascular disorders.

The American Heart Association 2006 expert consensus panel proposed the following definition for cardiomyopathy, which reflects the idea that many cardiomyopathies have an underlying etiology. Ischemia from CAD is probably the most common.

Cardiomyopathies are a heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilatation and are due to a variety of causes that frequently are genetic. Cardiomyopathies either are confined to the heart or are part of generalized systemic disorders, often leading to cardiovascular death or progressive heart failure-related disability.²¹²

The classification of cardiomyopathies is problematic. There was much confusion using the former classification of dilated, hypertrophic, and restrictive categories because of overlap when the same disease could appear in two different categories. And sometimes cardiomyopathy progresses from one category to another during the natural history of the disease, making classification difficult. As new knowledge of the pathogenesis of cardiomyopathy has unfolded and as new cardiomyopathies have been defined, the old classification scheme has been replaced with a new (but probably not final) classification.²¹²

Cardiomyopathies are classified as *primary* and *secondary*, based on predominant organ involvement. Primary cardiomyopathies include genetic, mixed (genetic and nongenetic), and acquired (Table 12-14). They are confined to the heart muscle.

Genetic cardiomyopathies include hypertrophic and arrhythmogenic right ventricular cardiomyopathies, left ventricular noncompaction, conduction system disease, and ion channelopathies. In general, these congenital or familial types of cardiomyopathies are fairly uncommon individually, but a growing number of different types caused by mutations in genetic encoding have been identified.

Mixed cardiomyopathies included dilated and primary restrictive nonhypertrophied cardiomyopathies. An example of an acquired cardiomyopathy is myocarditis. Considerable overlap can occur among the primary classifications within the same person (see Pathogenesis).

Secondary cardiomyopathies involve myocardial pathology as part of a large number and variety of generalized systemic disorders that affect the heart along with other organs at the same time.

Incidence and Risk Factors. Cardiomyopathy can affect any age group and is often seen in young adults in the second and third decades. The actual incidence is

Table 12-14 American Heart Association Classification of Cardiomyopathies

Primary*	Secondary†
Genetic <ul style="list-style-type: none"> Hypertrophic cardiomyopathy Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D; rare) Left ventricular noncompaction (LVNC) Conduction defects Ion channel disorders 	Infiltrative <ul style="list-style-type: none"> Amyloidosis Gaucher's disease (genetic/familial) Hurler's disease (genetic/familial) Hunter's disease (genetic/familial)
Mixed (genetic and nongenetic) <ul style="list-style-type: none"> Dilated cardiomyopathy Restrictive (nonhypertrophied; rare) 	Storage <ul style="list-style-type: none"> Hemochromatosis Glycogen storage disease Niemann-Pick disease (genetic/familial)
Acquired <ul style="list-style-type: none"> Myocarditis (inflammatory cardiomyopathy) Stress induced Peripartum/postpartum (rare) Cardiomyopathy in infants of mothers with insulin-dependent diabetes mellitus 	Toxicity <ul style="list-style-type: none"> Drugs, heavy metals, chemical agents
	Endomyocardial fibrosis
	Inflammatory (sarcoidosis)
	Endocrine <ul style="list-style-type: none"> Diabetes Hyperthyroidism Hypothyroidism Hyperparathyroidism Pheochromocytoma Acromegaly
	Neuromuscular/neurologic <ul style="list-style-type: none"> Friedreich's ataxia (genetic/familial) Muscular dystrophy (genetic/familial) Neurofibromatosis (genetic/familial) Tuberous sclerosis
	Nutritional deficiencies
	Autoimmune <ul style="list-style-type: none"> Systemic lupus erythematosus Dermatomyositis Rheumatoid arthritis Scleroderma Polyarteritis nodosa
	Electrolyte imbalance
	Cancer treatment (chemotherapy, radiation therapy)

Data from Maron BJ, Towbin JA, Thiene G, et al: Contemporary definitions and classification of the cardiomyopathies: an American Heart Association scientific statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation* 113(14):1807-1816, April 11, 2006.

*Predominantly involves the heart.

†Myocardial changes occur as part of a generalized systemic disorder affecting many organs; previously referred to as *specific cardiomyopathies*. Only the most common diseases associated with cardiomyopathies are listed.

unknown, but the disease may be more common than was previously realized.

This increase in incidence may be attributed to two important variables: (1) improved technology, which has allowed for more accurate evaluation of ventricular dimensions and ventricular wall movement; and (2) an increased incidence of myocarditis, an important precursor to cardiomyopathy, as a result of a wide variety of pathogens, toxins, and autoimmune reactions.

Delayed-onset cardiotoxic effects of chemotherapeutic agents may appear as chronic cardiomyopathy. Risk factors for the development of this type of cardiomyopathy include increasing doses of chemotherapeutic agents and previous mediastinal radiation.³⁴⁵ Doxorubicin hydrochloride (Adriamycin) and daunorubicin hydrochloride (Cerubidine) are the two agents recognized most often in association with dilated cardiomyopathy.

Dilated cardiomyopathy occurs most often in black men between the ages of 40 and 60 years. About one half of the cases of dilated cardiomyopathy are idiopathic, and the remainder result from some known disease process (e.g., rheumatic fever, myasthenia gravis, progressive muscular dystrophy, hemochromatosis, amyloidosis, sarcoidosis). Risk factors for dilated cardiomyopathy may include obesity, long-term alcohol abuse, systemic hypertension, cigarette smoking, infections, and pregnancy.

Peripartum cardiomyopathy is a rare but very serious disease that results in heart failure. It may appear for no apparent reason during the last month of pregnancy or shortly after delivery; incidence is higher among multiparous women older than 30 years, particularly those with malnutrition or preeclampsia. Estimates vary, but the occurrence may be 1 in every 1300 to 4000 deliveries. Maternal death from CHF, blood clots, and infection, and

stillbirth can occur. Symptoms of orthopnea, cough, palpitations, and high blood pressure may not occur until several weeks after delivery.

Hypertrophic cardiomyopathy appears to be genetically transmitted as an autosomal dominant trait on chromosome 14; currently 11 mutant genes have been linked with hypertrophic cardiomyopathy. It is still the most frequently occurring cardiomyopathy and the most common cause of sudden cardiac death in the young (including trained athletes).²¹²

Restrictive cardiomyopathy occurs as a result of myocardial fibrosis (e.g., amyloidosis, sarcoidosis, hemochromatosis), hypertrophy, infiltration, or defect in myocardial relaxation.

Pathogenesis. The exact pathogenesis of cardiomyopathy is unknown; the risk factors mentioned previously seem to lower the threshold for the development of cardiomyopathy. For example, heavy consumption of alcohol is thought to cause dilated cardiomyopathy through three mechanisms: direct toxic effect of alcohol or of its metabolites; effects of nutritional deficiencies, especially thiamine deficiency; and toxic effects of beverage additives, such as cobalt.

Obesity produces an increase in total blood volume and cardiac output because of the high metabolic activity of excessive fat. In moderate to severe cases of obesity, this may lead to left ventricular dilation, increased left ventricular wall stress, and left ventricular diastolic dysfunction.

Regardless of the underlying cause, dilated cardiomyopathy results from extensively damaged myocardial muscle fibers and is characterized by cardiac enlargement. The heart ejects blood less efficiently than normal, so that a large volume of blood remains in the left ventricle after systole, which results in ventricular dilation with enlargement and dilation of all four chambers and eventually leads to CHF (Figs. 12-16 and 12-17).

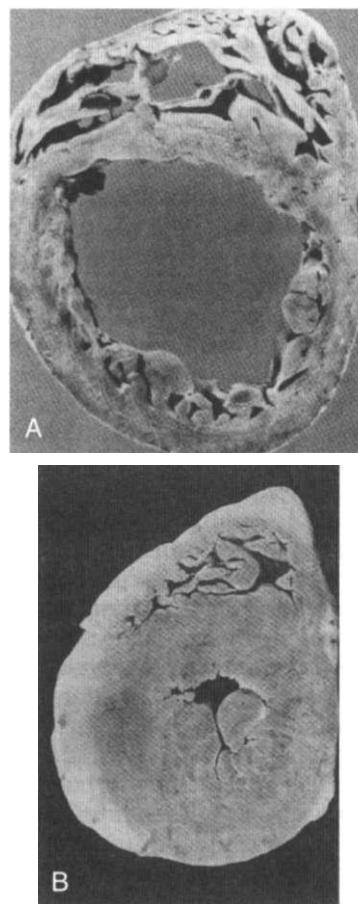


Figure 12-16

A, Cross-sectional view of dilated cardiomyopathy. **B**, Hypertrophied heart. (From Kinney M: Comprehensive cardiac care, ed 7, St Louis, 1991, Mosby, pp 346, 349.)

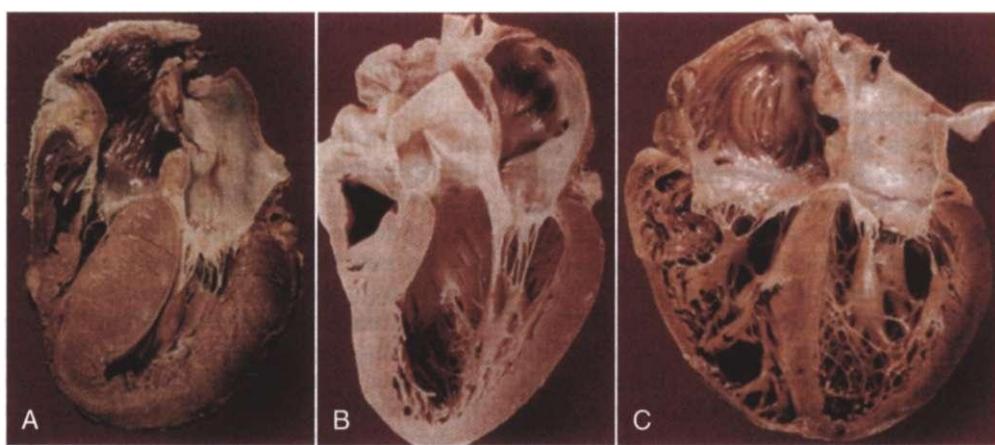


Figure 12-17

Gross pathologic specimens of the cardiomyopathies. **A**, Hypertrophic cardiomyopathy, showing a marked increase in myocardial mass and preferential hypertrophy of the interventricular septum. **B**, Normal heart, with normal left ventricular dimensions and thickness. **C**, Dilated cardiomyopathy, showing marked increase in chamber size. Atrial enlargement is also evident in both cardiomyopathies (**A** and **C**). (From Seidman JG, Seidman C: The genetic basis for cardiomyopathy: from mutation identification to mechanistic paradigms, *Cell* 104:557, 2001.)

Hypertrophic cardiomyopathy is distinguished by inappropriate and excessive left ventricular hypertrophy (thickening of the interventricular septum) and normal or even enhanced cardiac muscle contractile function. Over time, the overgrowth of the wall leads to rigidity in the myocardium. The result is decreased diastolic functioning, since the rigid myocardium cannot relax during the diastolic phase, reducing the amount of blood flowing into the ventricles. Restrictive cardiomyopathy is the least common form; it is identified by marked endocardial scarring (fibrosis) of the ventricles, and the resulting rigidity impairs diastolic filling.

Clinical Manifestations. Generally, the symptoms of cardiomyopathy are the same as for heart failure (e.g., dyspnea, orthopnea, tachycardia, palpitations, peripheral edema, distended jugular vein).

Dilated cardiomyopathy is characterized by fatigue and weakness; chest pain (unlike angina) may occur. Blood pressure is usually normal or low.

Hypertrophic cardiomyopathy is frequently asymptomatic, sudden death being the presenting sign; in fact, hypertrophic cardiomyopathy is the most common cause of sudden death in young competitive athletes. The most common symptom is dyspnea caused by high pulmonary pressures produced by the elevated left ventricular diastolic pressure; symptoms are often exacerbated during strenuous exercise.

Restrictive cardiomyopathy causes clinical manifestations related to decreasing cardiac output. As cardiac output falls and intraventricular pressures rise, signs of CHF appear. The earliest manifestations may include exercise intolerance, fatigue, and shortness of breath followed by other symptoms such as peripheral edema and ascites.

MEDICAL MANAGEMENT

DIAGNOSIS AND TREATMENT. Diagnosis requires exclusion of other causes of cardiac dysfunction, especially causes of CHF and arrhythmias. Catheterization to assess arteries and valves, echocardiography, chest radiography, blood chemistries, deoxyribonucleic acid (DNA) analysis (for hypertrophic cardiomyopathy), and ECG are specific tests performed. Researchers continue to investigate ways to monitor people with heart failure and to devise non-invasive diagnostic techniques.

The specific treatment of cardiomyopathy is determined by the underlying cause and may include physical, dietary, or pharmacologic interventions; mechanical circulatory support; or surgical intervention, including transplantation. Cardiac resynchronization therapy, the use of a pacemaker-like device to electrically stimulate both ventricles simultaneously (biventricular pacing), has been approved for use in CHF and is under investigation for use with dilated cardiomyopathy.

Alternatively, a cardiac support device called a "heart jacket" is under investigation for use in the United States for cardiomyopathy. This specially designed polyester material is stitched into place around the heart to prevent diseased heart muscle from further enlargement. Clinical safety trials are under way at the University of Pennsylvania.

Idiopathic dilated cardiomyopathy has no known cause; therefore there is no specific therapy. In contrast

to the other forms of cardiomyopathy, the progression of myocardial dysfunction in dilated cardiomyopathy may be stopped or reversed if alcohol consumption is reduced or stopped early in the course of the disease.

The β -blockers have an important immunoregulatory role in modifying the dysregulated cytokine network and reducing myocardial contractility and workload.²⁴⁵ Calcium channel blocking agents (see Table 12-5) may be used to relieve symptoms and reduce exercise intolerance. Restrictive cardiomyopathy has no specific treatment interventions. The goal is to control CHF through the use of diuretics, vasodilators, and salt restriction.

PROGNOSIS. Seventy-five percent of persons diagnosed with idiopathic dilated cardiomyopathy die within 5 years after the onset of symptoms, because diagnosis does not usually occur until advanced stages. Persons with hypertrophic cardiomyopathy can lead long, relatively asymptomatic lives; some people have a history of gradually progressive symptoms, but others experience sudden death, especially during exercise, as the initial diagnostic event. Restrictive cardiomyopathy may cause sudden death as a result of arrhythmia, or a more progressive course may occur, with eventual heart failure. Intervention rarely results in long-term improvement.

Many persons with various types of cardiomyopathy experience stabilization or even an improvement in symptoms, but the end result of cardiomyopathy is sudden death or a fatal progression toward heart failure. No cure exists, outside of cardiac transplantation. Heart transplantation shows a 1-year survival rate of over 80% and a 3-year survival rate of 70% for dilated cardiomyopathy. The 1-year survival rate without transplant is 5%.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-10

Cardiomyopathy

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

Other patterns (e.g., musculoskeletal) may apply depending on the chronicity of the condition and level of physical activity.

Sudden death can occur, but the incidence is rare. It occurs more often in younger people who have cardiomyopathy, and it may be avoided by eliminating strenuous exercise (e.g., running, competitive sports) when a diagnosis has been established. Rest improves cardiac function and reduces heart size.

During the early stages of the disease, many people find it difficult to accept activity restrictions and need encouragement to follow guidelines for activity restriction. Clients should avoid poorly tolerated activities; combine rest with activity; understand that physical stress and emotional stress exacerbate the disease; learn correct breathing techniques, since the Valsalva

maneuver decreases the inflow of venous blood and impairs outflow and should be avoided; and understand that alcohol depresses myocardial contractility and should be eliminated.

The therapist can provide valuable information regarding energy conservation techniques (see Box 9-8) to assist persons with continued independence in activities of daily living and possibly even with improvement of activity tolerance. This is especially true for the person awaiting a cardiac transplant. The therapist involved with athletes (of all ages) is advised to follow the American Heart Association's guidelines for preparticipation screening and identifying athletes at risk for sudden cardiac death.^{94,212,213}

Cardiomyopathy associated with cardiotoxicity following chemotherapy is often clinically silent because of the clients' low levels of physical activity. An evaluation to screen for potential cardiopulmonary dysfunction is essential with these clients.

The evaluation should include an assessment of current physical activity levels and exercise tolerance and monitoring of heart rate and rhythm, blood pressure, respiratory responses, and any other signs and symptoms of exercise intolerance (e.g., dyspnea, fatigue, light-headedness or dizziness, pallor, palpitations, chest discomfort).³⁴⁵ A scale that rates perceived exertion (see Table 12-13) is often useful during the evaluation and for establishing initial exercise guidelines toward improving endurance.

For the person who has been hospitalized and has not ambulated yet, the therapist will need to assess tolerance to activities in bed before ambulating. During activities, monitor pulse, oxygen saturation, respirations, blood pressure, and color. The heart rate, systolic blood pressure, and respiratory rate normally increase in proportion to the exercise (movement) intensity, whereas the diastolic blood pressure changes minimally (+/-10 mm Hg).

Improved activity tolerance may be demonstrated by minimal change in pulse or blood pressure during activities with minimal fatigue after the activity. Pulse, respirations, and blood pressure should return to a normal range within 3 minutes of the end of the activity. Discontinue any activity that results in chest pain, severe dyspnea, cyanosis, dizziness, hypotension, or sustained tachycardia.

Abnormal responses include either blunted or excessive rises in heart rate or systolic blood pressure, excessive increases in diastolic blood pressure or respiratory rate, a fall in systolic blood pressure with increasing activity, or increasing irregularity of the pulse. These signs may be the result of cardiopulmonary toxicity or simply the result of deconditioning. Increasing irregularity in the pulse with pairs or runs of faster beats or more than six isolated irregular beats per minute must be reported to the physician.³⁴⁵ If the person is receiving diuretics, monitor for signs of too-vigorous diuresis (e.g., muscle cramps, orthostatic hypotension). If the person becomes hypotensive, use a supine position with legs elevated to increase venous return and to ensure cerebral blood flow.

Trauma

Nonpenetrating

Any blunt chest trauma, which is especially common in steering wheel impact from an automobile accident, may produce myocardial contusion, resulting in myocardial hemorrhage with little if any myocardial scar once healing is complete. Large contusions may lead to myocardial scars, cardiac rupture, CHF, or formation of aneurysms.

The chest pain of myocardial contusion is similar to that of MI and is often confused with musculoskeletal pain from soft tissue consequences of chest trauma. Myocardial contusion is usually treated similarly to MI, with initial monitoring and subsequent progressive ambulation and cardiac rehabilitation (see Special Implications for the Therapist: Myocardial Infarction).

Penetrating

Penetrating cardiac injuries are most often due to external objects, such as bullets or knives, and sometimes from bony fragments secondary to chest injury. Iatrogenic causes of cardiac penetrating injury include perforation of the heart during catheterization and cardiac trauma from cardiopulmonary resuscitation. Complications include arrhythmias, aneurysm formation, death from infection (e.g., bacterial endocarditis or infection from a retained foreign body), a form of pericarditis associated with this type of injury, ventricular septal defects, and foreign body embolus.

Myocardial Neoplasm

Primary cardiac tumors are rare, with an autopsy frequency of 0.001% to 0.030%.⁵⁶ Malignant cardiac tumors account for 25% of primary cardiac tumors, with 95% of these tumors being sarcomas arising from connective tissue (e.g., angiosarcoma, rhabdomyosarcoma, mesothelioma, fibrosarcoma) and the remaining 5% being lymphomas.

Some of these sarcomas are limited to the myocardium, replacing functional cardiac tissue with cancerous cells without any intracavity extension. These tumors may produce no cardiac symptoms or may present with arrhythmias and conduction disturbances.

Tumors projecting into a cardiac cavity may present with progressive CHF, precordial pain, pericardial effusion tamponade, arrhythmias, conduction disturbances, and sudden death. Because these tumors occur more frequently in the right side of the heart, right-sided heart failure is more common (jugular venous distention, ascites, systemic edema). People with sarcomas face a rapid functional decline, with death occurring from a few weeks to 2 years after onset of symptoms. These tumors proliferate rapidly, invading and damaging not only the myocardium but contiguous structures such as the venae cavae and tricuspid valve as well.⁵⁶

Benign primary cardiac tumors occur approximately three times more often than malignant primary tumors, with myxomas accounting for nearly 50% of these primary benign tumors. Myxomas arise most often from the endothelial surface of the left atrium, causing mechanical interference with cardiac function including intracardiac

obstruction.⁴⁷ Tumors located in other cardiac chambers account for 10% of myxomas.⁵⁶ Other benign cardiac tumors (also rare) include lipoma, papilloma, fibroelastoma, rhabdomyoma, and fibroma.

Signs of obstruction can include right-sided heart failure, pulmonary edema, orthopnea, and dyspnea. Constitutional symptoms include fatigue, fever, weight loss, arthralgia, and myalgia. Embolization caused by fragments from the tumor can also occur in these individuals. If the tumor is in the left side of the heart, the emboli result in infarction damage to the viscera, including the heart, limbs, kidneys, and CNS.²⁷⁴ Because these tumors often lie in the atrial cavity they can (if large enough) cause damage to the mitral valve or even block the orifice of this valve, leading to sudden death. Tumors found in the right side of the heart infrequently lead to pulmonary hypertension and PEs.

Metastases to the heart and pericardium are much more common, occurring 100 to 1000 times more often than primary cardiac tumors.^{56,284} Melanoma has the highest frequency of metastasis to the heart, with metastases also possible from carcinomas of the lung, breast, and esophagus and malignant leukemia and lymphoma.²⁷⁵

Tumor may involve the heart by one of four metastatic pathways: retrograde lymphatic extension, hematogenous spread, direct contiguous extension, or transvenous extension. Metastatic involvement of the heart and pericardium may go unrecognized until autopsy. Impairment of cardiac function occurs in approximately 30% of cases and is usually attributed to pericardial effusion. The clinical presentation includes shortness of breath, cough, anterior thoracic pain, pleuritic chest pain, or peripheral edema. Cardiac neoplasms come to the attention of a therapist when (1) progressive interference with mitral valve function results in exercise intolerance or exertional dyspnea; (2) embolus causes a stroke; or (3) systemic manifestations occur, including muscle atrophy, arthralgias, malaise, or Raynaud's phenomenon.

Diagnosis of myxomas and other cardiac neoplasms is usually made by echocardiography followed by imaging studies, with MRI being of greater value in delineating cardiac tumors.²⁸⁴ There are no specific physical or laboratory tests for metastatic heart disease, and diagnosis is difficult as these tumors can masquerade as other cardiac defects. ECG is nonspecific, chest radiography may reveal an enlarged cardiac silhouette, and radionuclide angiography is helpful in diagnosing intracavity tumors. Two-dimensional echocardiography is the method of choice to detect cardiac metastases.²⁷⁵

Treatment of choice for myxomas is usually resection of the tumor, which in most cases is curative. Cardiac rehabilitation may be required according to the individual's postoperative cardiovascular condition. Recurrence is rare and appears to be the result of incomplete resection of the tumor or intraoperative dislocation of tumor material. The presence of cancer cells in more than one area of the myocardium (multifocal genesis) may also lead to recurrence despite treatment.²⁷⁴

In most cases, cardiac metastases are treated with palliative care because in most cases, advanced disease is present at the time of diagnosis. Radiation is not typically

used to treat cardiac neoplasms, which means that radiation heart disease occurs secondarily to radiation therapy for tumors in the area of the heart (e.g., mediastinum, breast, head and neck, and thyroid). A history of such tumors should alert the therapist to the possibility that cardiac defects may be present.

Congenital Heart Disease

Overview and Incidence

Congenital heart disease is an anatomic defect in the heart that develops in utero during the first trimester and is present at birth. Over the past three decades, major advances have been made in the diagnosis and treatment of congenital heart disease, resulting in many more children who have survived to adulthood with surgically corrected or uncorrected anomalies. Today, there are over 1 million adults with congenital heart conditions. Congenital heart disease affects about 8 of every 1000 babies born in the United States, making this the most common category of congenital structural malformation. Other than prematurity, it is the major cause of death in the first year of life. Children with congenital heart disease are also more likely to have extracardiac defects, such as tracheoesophageal fistula, diaphragmatic hernias, and renal abnormalities.

There are two categories of congenital heart disease: cyanotic and acyanotic (Table 12-15). In clinical practice this system of classification is problematic, because children with acyanotic defects may develop cyanosis and those with cyanotic defects may be pink and have more clinical signs of CHF.

Cyanotic defects result from obstruction of blood flow to the lungs or mixing of desaturated blue venous blood with fully saturated red arterial blood within the chambers of the heart. Most *acyanotic* defects involve primarily left-to-right shunting through an abnormal opening.

Etiologic Factors

Many congenital heart diseases have genetic causes with well-known chromosomal anomalies (e.g., trisomy 13, 18, 21; Turner's syndrome). Approximately 10% of all congenital heart defects are known to be associated with a single identified mutant gene or chromosomal abnormalities; for the remainder, the causes are either unknown or involve multiple factors, such as diabetes, alcohol consumption, viruses, maternal rubella infection during the first trimester, and drugs such as thalidomide.

In the case of atrial septal defect, most result from spontaneous genetic mutations, although some are inherited. Patent ductus arteriosus occurs in pregnancies complicated by persistent perinatal hypoxemia or maternal rubella infection and among infants born at high altitude or prematurely.⁵⁰

Pathogenesis

The heart begins to form from a tubelike structure during the fourth week after conception. As development progresses, the tube lengthens and forms chambers, septa, and valves. Anything that interferes with this developmental process during the first 8 to 10 weeks of pregnancy can result in a congenital defect (Fig. 12-18).

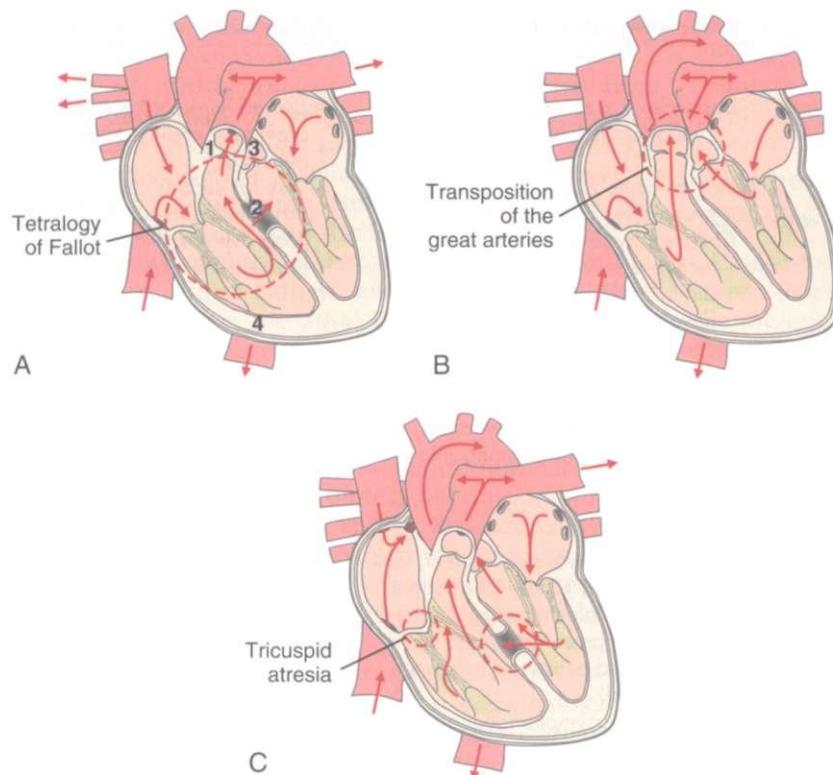
Table 12-15 Congenital Heart Disease

Defect	Incidence	Clinical Manifestations	Prognosis
Cyanotic			
Transposition of the great vessels	16%*; 3:1 male/female ratio	Depends on size and type of defects; cyanosis; CHF (newborn)	Improved surgical treatment provides excellent long-term outcome
Tetralogy of Fallot	10%-15%	<i>Infants:</i> acutely cyanotic at birth or progressive cyanosis first year <i>Children:</i> hypoxic events with tachypnea, increasing cyanosis, digital clubbing; poor growth and development; seizures, loss of consciousness, death possible <i>Adults:</i> dyspnea, limited exercise tolerance Newborn cyanosis; tachycardia; dyspnea; digital clubbing (older child)	At risk for sudden lethal arrhythmias; mild obstruction progresses with age; reduced life expectancy
Tricuspid atresia	<1%; relatively rare		Unreported; depends on success of treatment
Acyanotic			
Ventricular septal defect	25%; single most common CHD; 25%-40% close spontaneously by age 2 yr; 90% close by age 10 yr	Asymptomatic with small defect; CHF (age 1-6 mo); history of frequent respiratory infections; poor growth and development; dyspnea, fatigue, and exercise intolerance (older child)	No physical restrictions
Atrial septal defect	10%; 2:1 female/male ratio Accounts for 33% of all CHD cases surviving to adulthood	<i>Older child:</i> asymptomatic; growth failure; CHF <i>Adult:</i> fatigue or dyspnea on exertion	No physical restrictions if corrected; frequent complications in adults
Coarctation of the aorta	6%; 3:1 male/female ratio	High systolic BP and bounding pulses in arms; weak or absent femoral pulses; cool lower extremities with lower BP <i>Infants:</i> CHF <i>Children:</i> headaches, fainting, epistaxis (hypertension); exercise intolerance, easy fatigability <i>Adults:</i> asymptomatic or signs of hypertension (headache, epistaxis, dizziness, palpitations) <i>Children:</i> asymptomatic; CHF <i>Adult:</i> if symptomatic: fatigue, dyspnea, palpitations	Good if survive to childhood; exercise testing recommended before participation in athletics; reduced life expectancy; increased risk of aortic dissection during pregnancy
Patent ductus arteriosus	12%; spontaneous closure in normal-term infants by day 4; common in children born to mothers affected by rubella during first trimester; increased incidence in infants born at high altitudes (over 10,000 ft); present in 20%-60% of premature infants weighing <1500 g		Closure may occur up to age 2 yr; normal life expectancy with small defect; aneurysm and rupture can occur; poor prognosis for large defect without transplantation
Aortic stenosis	5% of all congenital heart disease	Asymptomatic; exercise intolerance, dizziness, and chest pain with prolonged standing	Good with early detection and surgical treatment; exercise testing recommended before participation in athletics

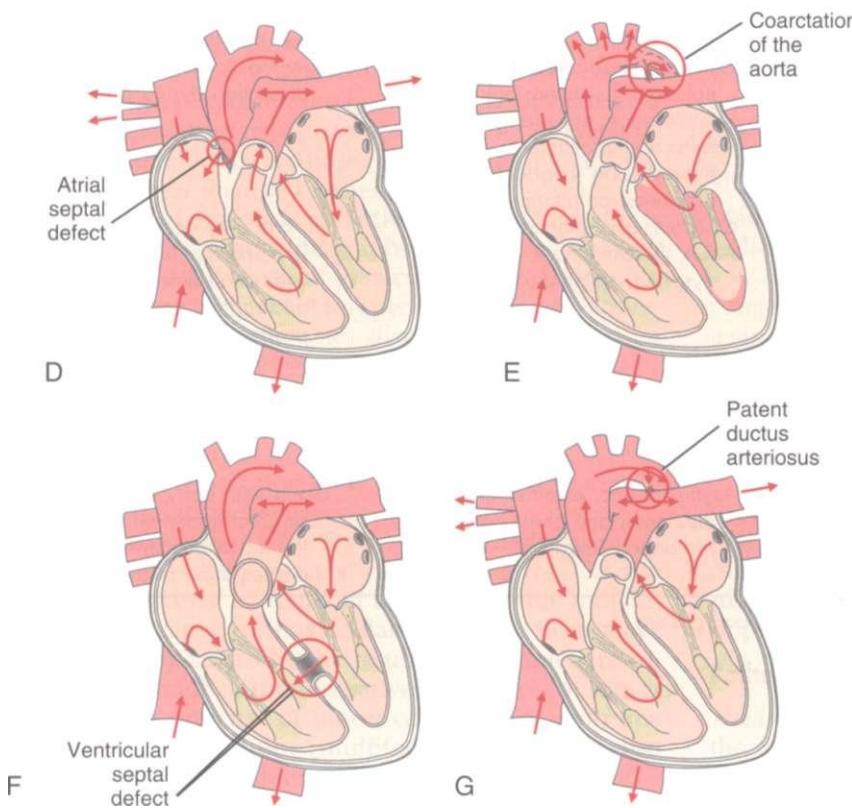
CHF, Congestive heart failure; CHD, congenital heart disease; BP, blood pressure.

*Figures represent percentage of all congenital heart disease.

MAJOR CYANOTIC DEFECTS



MAJOR ACYANOTIC DEFECTS

**Figure 12-18**

Major cyanotic defects (see Fig. 12-1 for normal structure and circulation of the heart): **A**, *Tetralogy of Fallot* has four defects: (1) pulmonary stenosis: narrowing at or just below the pulmonary valve; (2) ventricular septal defect (VSD): hole between the two bottom chambers (ventricles) of the heart; (3) aorta is positioned over the ventricular septal defect instead of in the left ventricle; (4) right ventricle is more muscular than normal. **B**, *Transposition of the great arteries*: systemic venous blood returns to the right atrium and then goes to the right ventricle and on to the aorta instead of going to the lung via the pulmonary artery. **C**, *Tricuspid atresia*: failure of the tricuspid valve to develop with a lack of communication from the right atrium to the right ventricle. Major acyanotic defects: **D**, *Atrial septal defect*: blood from the pulmonary vein enters the left atrium, and some blood crosses the atrial septal defect into the right atrium and ventricle. **E**, *Coarctation of the aorta*: severe obstruction of blood flow in the descending thoracic aorta. **F**, *Ventricular septal defect*: when the left ventricle contracts, it ejects some blood into the aorta and some across the ventricular septal defect into the right ventricle and pulmonary artery. **G**, *Patent ductus arteriosus*: some of the blood from the aorta crosses the ductus arteriosus and flows into the pulmonary artery.

Cyanotic. In *transposition of the great vessels (TGV)*, no communication exists between systemic and pulmonary circulations, so that the pulmonary artery leaves the left ventricle and the aorta exits from the right ventricle. In order for the infant with this condition to survive, there must be communication between the two circuits. In approximately one third of all cases, another associated defect occurs that permits intracardiac mixing (e.g., atrial septal defect, ventricular septal defect, patent ductus arteriosus), but two thirds have no other defect present and severe cyanosis develops.⁵¹

Tetralogy of Fallot consists of four classic defects: (1) pulmonary stenosis, (2) large ventricular septal defect, (3) aortic communication with both ventricles, and (4) right ventricular hypertrophy. *Tricuspid atresia* is a failure of the tricuspid valve to develop, with a lack of communication from the right atrium to the right ventricle. Blood flows through an atrial septal defect or a ductus arteriosus to the left side of the heart and through a ventricular septal defect to the right ventricle and out to the lungs. There is complete mixing of unoxygenated and oxygenated blood in the left side of the heart, resulting in systemic desaturation and varying amounts of pulmonary obstruction.

Acyanotic. *Ventricular septal defect* is an abnormal opening between the right and left ventricles that may vary in size from a small pinhole to complete absence of the septum, resulting in a common ventricle. *Atrial septal defect* is an abnormal opening between the atria, allowing blood from the higher-pressure left atrium to flow into the lower-pressure right atrium.

Coarctation of the aorta is a localized narrowing near the insertion of the ductus arteriosus, resulting in increased pressure proximal to the defect (head, upper extremities) and decreased pressure distal to the obstruction (body, lower extremities).

Patent ductus arteriosus is a failure of the fetal ductus arteriosus (artery connecting the aorta and pulmonary artery) to close within the first weeks of life. The continued function of this vessel allows blood to flow from the high-pressure aorta to the low-pressure pulmonary artery, causing continuous flow from the aorta to the pulmonary artery (referred to as left-to-right shunting). A patent ductus arteriosus rarely closes spontaneously after infancy.

Aortic stenosis is discussed later in this chapter in the section on Diseases Affecting the Heart Valves.

Clinical Manifestations

The most common signs and symptoms include cyanosis and signs of CHF (e.g., dyspnea, pulmonary edema, fatigue). See Table 12-15 for clinical manifestations of each particular defect. Complications may include heart failure, pulmonary edema, pneumonia, hypoxia, and sudden death. There is often a risk of bacterial endocarditis and pulmonary vascular obstructive disease later in life.

MEDICAL MANAGEMENT

PREVENTION AND DIAGNOSIS. As whole genome sequencing continues to develop, identification of genetic mutations predisposing to congenital heart disease may allow

preventive measures by modulation of secondary genetic or environmental factors.^{51,52} Until then, most forms of congenital heart disease can potentially be detected *in utero* with the routine use of ultrasonography.

The prenatal diagnosis of a major cardiac malformation requires further assessment for extracardiac and chromosomal disorders. Conversely, diagnosis of Down syndrome (prenatally or postnatally) requires early cardiologic assessment for cardiac anomalies, most commonly atrioventricular and ventricular septal defects. Prenatal knowledge of cardiac anomalies allows for optimal perinatal and postnatal management.

Prenatal screening for maternal rubella antibodies provides important information for further diagnostic testing. In cases where prenatal diagnosis does not occur and when there are no symptoms initially, cardiac anomalies can remain undetected for years and even decades. For example, a person with atrial septal defect may have normal sinus rhythm for the first three decades of life and then develop atrial fibrillation (AF) and supraventricular tachycardia (SVT).⁵¹ Clinical diagnosis begins with detection of signs and symptoms, auscultation, and detection of heart murmur. Transesophageal echocardiography, Doppler color-flow echocardiography and now RT-3D echocardiography provide a definitive diagnosis without invasive cardiac catheterization and angiography.

TREATMENT AND PROGNOSIS. Remarkable innovations in medical and surgical approaches over the past several decades now allow for correction of major cardiac defects in children, even in early infancy. Prenatal (*in utero*) correction has not been accomplished as yet. Postnatally, curative or palliative (providing relief of symptoms) surgical correction is now available for more than 90% of persons with congenital heart disease.

There is a clear trend toward complete correction of malformations rather than staged procedures to obtain initial palliation and delayed correction. The risk for most surgical procedures is low (between 1% and 5%). Gene transfer to create a patent ductus arteriosus in animal studies may lead the way for additional gene transfer techniques to be successful in humans in the future.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-11

Congenital Heart Disease

PREFERRED PRACTICE PATTERNS

5B: Impaired Neuromotor Development

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

Other patterns may apply depending on complications (e.g., pneumonia, heart failure).

Therapists need to be alert to signs of CHF in children with congenital heart disease and in infants with suspected congenital heart disease. Signs of CHF indicate a worsening clinical condition; the earlier these are detected, the sooner intervention can be initiated. (See also Special Implications for the Therapist: Congestive Heart Failure).

Continued.

The surgical procedures associated with the repair of congenital heart disease (e.g., bypass, deep hypothermia) are associated with an increased incidence of neurologic abnormalities. Neurodevelopmental deficits resulting from surgical repair of cardiac defects may include choreoathetosis, cerebral palsy, or hemiparesis.¹²³

Gross motor development can be negatively impacted by prolonged hospitalization, deficiencies in cardiovascular status, surgical techniques used to minimize blood loss, or any combination of these factors. The more complex the defect or defects and the more numerous the open heart surgeries required, the greater the risk for neurologic impairment.³²⁶

Most children with significant heart defects will have had heart surgery before they start school. In addition to a developmental assessment, the therapist should evaluate for soft tissue restriction at the site of the healed scar (either sternal or thoracic), which may affect breathing capacity.

Physiologic response to therapy intervention can be assessed by observing skin color, respiratory effort, and behavioral response. Oxygen saturation monitors may not be helpful, because these children have abnormally low readings as their baseline level.³²⁶ Data on exercise capacity after specific types of surgical procedures are available.³¹⁰

In general, anyone who has had successful surgery is allowed unrestricted sports activity. Young children with unrepaired tetralogy of Fallot instinctively learn to squat, getting into the flat-footed baseball catcher's stance when they are fatigued. This posture increases the tension in the leg muscles, reduces blood flow to the leg muscles, and raises peripheral resistance and blood pressure.

Twenty or thirty years ago, diagnosis of congenital defects was much more difficult, and many anomalies went undetected. Adults with undiagnosed congenital defects may develop exercise intolerance, shortness of breath, palpitations, blood pressure irregularities, or symptoms of CHF, which should alert the therapist to the need for medical referral.

Exercise recommendations for children and adolescents with congenital malformations are available.⁹⁴ Care of pregnant women with congenital heart disease requires understanding of the specific congenital defect, the nature of previous surgical correction, and the presence of any complications or sequelae.⁸¹

portion of the heart. The signal from the SA node travels through the cardiac conduction system, first through the walls of the atria and then through the walls of the ventricles, causing the atrial (supraventricular) and ventricular chambers of the heart to contract and relax at regular rates necessary to maintain circulation at different levels of activity. An arrhythmia (dysrhythmia) is a disturbance of heart rate or rhythm caused by an abnormal rate of electrical impulse generation by the SA node or the abnormal conduction of impulses.

Arrhythmias can be classified according to their origin as ventricular or supraventricular (atrial), according to the pattern (fibrillation or flutter), or according to the speed or rate at which they occur (tachycardia or bradycardia).

Several types of AF are now recognized, including first-detected-episode AF (may or may not be symptomatic; may self-resolve), recurrent paroxysmal AF (two or more episodes that resolve spontaneously), persistent AF, and permanent AF. Persistent AF is sustained for more than 7 days. It can occur after a first-detected-episode AF or after recurrent paroxysmal AF. Permanent AF, also known as chronic AF, occurs when sinus rhythm cannot be sustained after cardioversion (normal heart rhythm returns spontaneously) or when the decision has been made to let AF continue without efforts to restore normal sinus rhythm.¹¹⁷

Arrhythmias vary in severity from mild, asymptomatic disturbances that require no intervention (e.g., sinus arrhythmia, in which the heart rate increases and decreases with respiration) to catastrophic ventricular fibrillation, which requires immediate resuscitation. The clinical significance depends on the effect on cardiac output and blood pressure, which is partially influenced by the site of origin.

Etiologic Factors and Incidence

Arrhythmias may be congenital or may result from one of several factors, including hypertrophy of heart muscle fibers secondary to hypertension, previous MI, valvular heart disease, or degeneration of conductive tissue that is necessary to maintain normal heart rhythm (called *sick sinus syndrome*).

Chronic alcohol use and binge drinking have been linked with disturbances in cardiac rhythm, even in individuals without underlying heart disease. *Holiday heart syndrome* is the term used to describe acute arrhythmia (usually SVT) triggered by excessive alcohol intake in an otherwise healthy person. The affected individual experiences intermittent or continuous palpitations with dyspnea, dizziness, or chest pain often mentioned.

The prevalence of AF doubles with each advancing decade of age beginning at age 50 to 59 years, with a statistically significant increase among men age 65 to 84 years, although this gap closes with advancing age and remains unexplained.^{175,221}

BMI appears to correlate strongly with the risk of AF.¹⁰⁰ With each unit increment of BMI the risk of AF increases 3%. A person who is obese has about a 34% greater risk of AF when compared to a person with normal BMI. Moreover, people in the heaviest BMI category have 2.3 times the risk. Improved cardiac care has increased the

DISEASE AFFECTING THE CARDIAC NERVOUS SYSTEM

Arrhythmias: Disturbances of Rate or Rhythm

Definition and Overview

The number of times the heart beats (rate) and the heart rhythm are generated and regulated by the sinoatrial (SA) node, the internal pacemaker located in the upper right

number of survivors of cardiac incidents who may experience subsequent complications, such as arrhythmias.

Cardiac arrhythmias are very common in the setting of heart failure, with atrial and ventricular arrhythmias often present in the same person. Arrhythmias can occur when a portion of the heart is temporarily deprived of oxygen, disturbing the normal pathway of the heartbeat. Toxic doses of cardioactive drugs (e.g., digoxin and other cardiac glycosides), phenylpropanolamine found in some decongestants, alcohol and caffeine consumption, high fevers, and excessive production of thyroid hormone (hyperthyroidism) may also lead to arrhythmias. In many cases, particularly in younger people, there is no known or apparent cause.

Pathogenesis and Clinical Manifestations

Rate. The adult heart beats an average of 60 to 100 beats/min; an arrhythmia is considered to be any significant deviation from the normal range. Whether change in heart rate (number of contractions of the cardiac ventricles per period of time) produces symptoms at rest or on exertion depends on the underlying state of the cardiac muscle and its ability to alter its stroke output to compensate.

Rate arrhythmias are of two basic types: tachycardia and bradycardia. Tachycardia occurs when the heart beats too fast (more than 100 beats/min). Tachycardia develops in the presence of increased sympathetic stimulation, such as occurs with fear, pain, emotion, exertion, or exercise; or with ingestion of artificial stimulants, such as caffeine, nicotine, and amphetamines.

Tachycardia is also found in situations in which the demands for oxygen are increased, such as fever, CHF, infection, anemia, hemorrhage, myocardial injury, and hyperthyroidism. Usually the individual with tachycardia perceives no symptoms, and medical intervention is directed toward the underlying cause.

Bradycardia (less than 50 beats/min) is normal in well-trained athletes, but it is also common in individuals taking P-blockers, those who have had traumatic brain injuries or brain tumors, and those experiencing increased vagal stimulation (e.g., from suctioning or vomiting) to the physiologic pacemaker.

Organic disease of the sinus node, especially in older people and those with heart disease, can also cause sinus bradycardia. Bradycardia is usually asymptomatic, but when it is caused by a pathologic condition, the person may experience fatigue, dyspnea, syncope, dizziness, angina, or diaphoresis (profuse perspiration). Medical intervention is not usually required unless symptoms interfere with function or are drug or angina induced; atropine or a mechanical pacemaker can be used to reestablish a more normal heart rate.

Rhythm. Arrhythmias as variations from the normal rhythm of the heart (especially the heartbeat) are detected when they become symptomatic or during monitoring for another cardiac condition. Abnormalities of cardiac rhythm and electrical conduction can be lethal (sudden cardiac death), symptomatic (syncope or near syncope, dizziness, chest pain, dyspnea, palpitations), or asymptomatic. They are dangerous because they reduce cardiac output so that perfusion of the brain or myocardium is

impaired, or they tend to deteriorate into more serious arrhythmias with the same consequences.

The many different types of abnormal cardiac rhythms are usually classified according to their origin (atrial, ventricular), but only the most common ones are included here. Complete discussion of all other cardiac arrhythmias is available.¹⁵⁰

Sinus arrhythmia is an irregularity in rhythm that may be a normal variation in athletes, children, and older people or may be caused by an alteration in vagal stimulation. Sinus arrhythmia may be respiratory (increases and decreases with respiration) or nonrespiratory and associated with infection, drug toxicity (e.g., digoxin, morphine), or fever. Treatment for the respiratory type of sinus arrhythmia is not necessary; all other sinus arrhythmias are treated by providing intervention for the underlying cause.

AF is the most common type of SVT or chronic arrhythmia. SVT is also called paroxysmal supraventricular tachycardia (PSVT) or paroxysmal atrial tachycardia (PAT). It is characterized by rapid, involuntary, irregular muscular contractions of the atrial myocardium—quivering or fluttering instead of contracting normally. Consequently, blood remains in the atria after they contract and the ventricles do not fill properly. The heart races, but blood flow may diminish, creating a drop in oxygen levels that results in symptoms of shortness of breath, palpitations, fatigue, and, more rarely, fainting. AF occurs most often as a secondary arrhythmia associated with rheumatic heart disease, dilated cardiomyopathy, atrial septal defect, hypertension, mitral valve prolapse, recurrent cardiac surgery, and hypertrophic cardiomyopathy (conditions that affect the atria).

Secondary AF can also occur in people without cardiac disease but in the presence of a systemic abnormality that predisposes the individual to arrhythmia (e.g., hyperthyroidism, medications, diabetes, obesity, pneumonia, or alcohol intoxication or withdrawal). People with AF are prone to blood clots because blood components that remain in the atria aggregate and attract other components, triggering clot formation. The effect rarely occurs before 72 hours of the first abnormal contraction. AF can result in CHF, cardiac ischemia, and arterial emboli that can result in an ischemic stroke.

Ventricular fibrillation is an electrical phenomenon that results in involuntary uncoordinated muscular contractions of the ventricular muscle; it is a frequent cause of cardiac arrest. Treatment is directed toward depolarizing the muscle, thus ending the irregular contractions and allowing the heart to resume normal regular contractions.

Heart block is a disorder of the heartbeat caused by an interruption in the passage of impulses through the heart's electrical system. This may occur because the SA node misfires or the impulses it generates are not properly transmitted through the heart's conduction system. Heart blocks are differentiated into three types determined by ECG testing: first-degree, second-degree, and third-degree (complete) heart block. Causes include CAD, hypertension, myocarditis, and overdose of cardiac medications (e.g., digitalis, calcium channel blockers, β -blockers). Depending on the degree of the heart block, it

can cause fatigue, dizziness, or fainting. Heart block can affect people at any age, but this condition primarily affects older people. Mild cases do not require intervention; medication and pacemakers are the two primary forms of management for symptomatic cases.

Sick sinus syndrome, or brady-tachy syndrome, is a complex cardiac arrhythmia and conduction disturbance that is associated with advanced age, CAD, or drug therapy (e.g., digitalis, calcium channel blockers, β -blockers, antiarrhythmics). Sick sinus syndrome as a result of degeneration of conductive tissue necessary to maintain normal heart rhythm occurs most often among older people. A variety of other heart diseases and other conditions (e.g., cardiomyopathy, sarcoidosis, amyloidosis) also may result in sinus node dysfunction. Sick sinus syndrome is characterized by bradycardia alone, bradycardia alternating with tachycardia, or bradycardia with atrioventricular block resulting in cerebral manifestations of light-headedness, dizziness, and near or true syncope.

Sinus node dysfunction is suspected in the older adult experiencing episodes of syncope or near syncope, especially in the presence of heart palpitations. An accurate diagnosis is made with ECG, often requiring a 24-hour Holter monitor to document the arrhythmias described. Treatment for the symptomatic person varies according to the specific arrhythmia manifestations and may include antiarrhythmic agents alone or combined with a permanent-demand pacemaker or withdrawal of agents that may be responsible.

Holiday heart syndrome may occur when the heart responds to the increase in catecholamines (epinephrine, norepinephrine) brought on by excessive alcohol intake. Alcohol metabolites may also cause conduction delays. The toxic effects of alcohol can also cause a rise in the level of free fatty acids, contributing to the onset of this condition.

MEDICAL MANAGEMENT

DIAGNOSIS. ECG is the most common test procedure to document arrhythmias, but if the person is not experiencing symptoms, the heartbeat may look normal. Tape-recorded ambulatory ECG may be used to document arrhythmias. The individual may use continuous monitoring (external cardiac monitoring; Holter monitoring; Fig. 12-19) recording all cardiac cycles over a prescribed period of time (usually 24 to 48 hours) or cardiac event monitoring recording ECG just when symptoms are perceived.

Monitoring is especially helpful in recording sporadic arrhythmias that an office or stress test ECG might miss. Monitoring may also be used by persons recovering from MIs, receiving antiarrhythmic medications, or using pacemakers. New pocket-sized devices to allow home monitoring are available; readings may be stored, and the device can be hooked up to the physician's ECG or diagnostic computer or transmitted over the telephone. For symptoms that occur rarely (e.g., once every 6 months), an insertable loop recorder can be used. This small device is implanted under the skin in the chest using a local anesthetic. Monitoring units do not replace an ECG and should not be used without a physician's approval.

TEE imaging study using an ultrasonic transducer mounted on the tip of a flexible instrument is used to detect cardiac emboli before medications are initiated to control rate and rhythm. If a serious arrhythmia is suspected, an electrophysiologic study (EPS) can be performed. This test is an invasive study that uses wires placed via catheterization to electronically stimulate the heart in an attempt to reproduce the arrhythmia.

TREATMENT. The goal of treatment is to control ventricular rate, prevent thromboembolism, and restore normal sinus rhythm if possible. Normal heart rhythm returns spontaneously (called *cardioversion*) almost immediately in some cases, especially if there is no underlying heart disease. When conversion to normal rate and rhythm does not occur, there are two major approaches to cardioversion: electrical and pharmacologic.

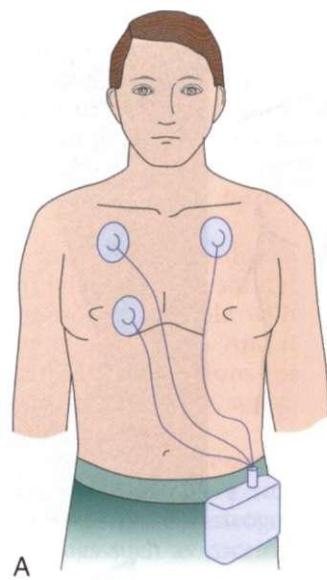
The electrical method employs the use of a device called a *defibrillator* and is usually most effective and may require several weeks of anticoagulant therapy (warfarin) to reduce stroke risk. Anyone who has been in AF less than 48 hours but is hemodynamically unstable with serious signs and symptoms related to AF will need immediate electrical cardioversion. Low-voltage electric shocks interrupt the irritable foci of the heart, letting the SA node resume its role as a primary pacemaker.²⁸⁵

Pharmacologic treatment may include agents prolonging depolarization and/or other cardiovascular medications (see Table 12-5). If successful, cardioversion restores sinus rhythm, and drug therapy is used to maintain normal heart rate and rhythm. Even with successful electrical cardioversion, long-term antiarrhythmic and anti-coagulation drug therapy is used to sustain normal sinus rhythm.

Some tachycardias can be treated with radio-wave ablation, a nonsurgical but invasive technique that uses catheterization to thread wires into the heart through which radio waves can be aimed at the heart tissue where the arrhythmia originates. The catheter-delivered quick bursts of current destroy the specific areas of heart muscle that are generating the abnormal electrical signals causing the arrhythmia. One complication of this technique is the potential destruction of the conducting system (the heart's own internal pacemaker), which necessitates surgical implantation of an artificial pacemaker for some people.

Pacemakers, implants designed to replace the heartbeat by delivering a battery-supplied electrical stimulus through leads attached to electrodes in contact with the heart, may be used in cases of bradycardia, heart block, or refractory tachycardia. Refractory tachycardia is a condition in which the heart is beating very quickly, but only a portion of those beats are functional; many more beats just echo or make a beat but without contractile force behind the blood flow. Functionally, the heartbeat is actually very slow.

Pacemakers initiate the heartbeat when the heart's intrinsic conduction system fails or is unreliable. In the case of life-threatening arrhythmias (e.g., ventricular tachycardia, ventricular fibrillation) that do not respond to other types of intervention, a device called an implant-



A



B



C

Figure 12-19

A, External cardiac monitoring (a form of telemetry, also called ambulatory electrocardiography [ECG] or Holter monitoring) uses a tape recorder that is attached to the skin by ECG electrodes. It is able to record the heart rhythm over a 24-hour period. Any symptoms experienced while wearing the unit should be recorded by the individual wearing the device. The recording is then analyzed. It may detect changes in heart rhythm or changes in the ECG that might indicate a lack of blood supply to the heart. **B,** Any number of electrodes up to 12 leads can be used. The standard three-electrode system in **A** consists of positive electrode, negative electrode, and ground electrode. **C,** The unit is small and convenient and can be clipped to the belt or waistband or slipped into a pocket. (Courtesy Cardiac Science Corporation, Bothell, WA. Used with permission.)

able cardioverter-defibrillator may be implanted (Fig. 12-20). The cardioverter-defibrillator monitors the heart rhythm, and if the heart starts beating abnormally, it generates an electric shock to restore the normal sinus (heart) rhythm.

For people whose arrhythmias are resistant to pharmacologic therapy, another surgical intervention is available called the *maze procedure*. This procedure requires open heart surgery and involves a series of mazelike cuts made in the atria and then sewn back together. The scar tissue that forms during the healing process blocks faulty circuits, preventing AF. Many people still need a pacemaker and drug therapy to maintain normal rate and rhythm. A more refined version of this procedure (catheter maze) takes a percutaneous, nonsurgical, noninvasive approach using radiofrequency ablation to destroy tissue.

A more recently developed treatment intervention called *ventricular resynchronization therapy* is gaining recognition for the treatment of intraventricular conduction disturbances associated with CHF. This redesigned pacemaker resynchronizes the right and left ventricles so they

pump at the same time, making the heart pump more forcefully instead of pumping faster (as occurs with a typical pacemaker or in the case of CHF when the heart beats faster to compensate for a weak pumping mechanism).³³⁹

PROGNOSIS. About half of all individuals with AF will spontaneously convert to normal sinus rhythm within 24 to 48 hours; this is less likely to occur in people whose AF has lasted more than 7 days.³⁶⁰

Sudden cardiac arrest (sudden death) is responsible for 300,000 deaths annually and is often preceded by fatal heart dysrhythmias in people who have no prior history of heart disease. In fact, new data from the Framingham Heart Study indicate that AF is independently associated with a substantially increased risk for death in both men and women, even after adjustment for age and associated factors, such as hypertension, CHF, and stroke.

Defibrillation within the first few minutes of cardiac arrest can save up to 50% of lives; by comparison, an

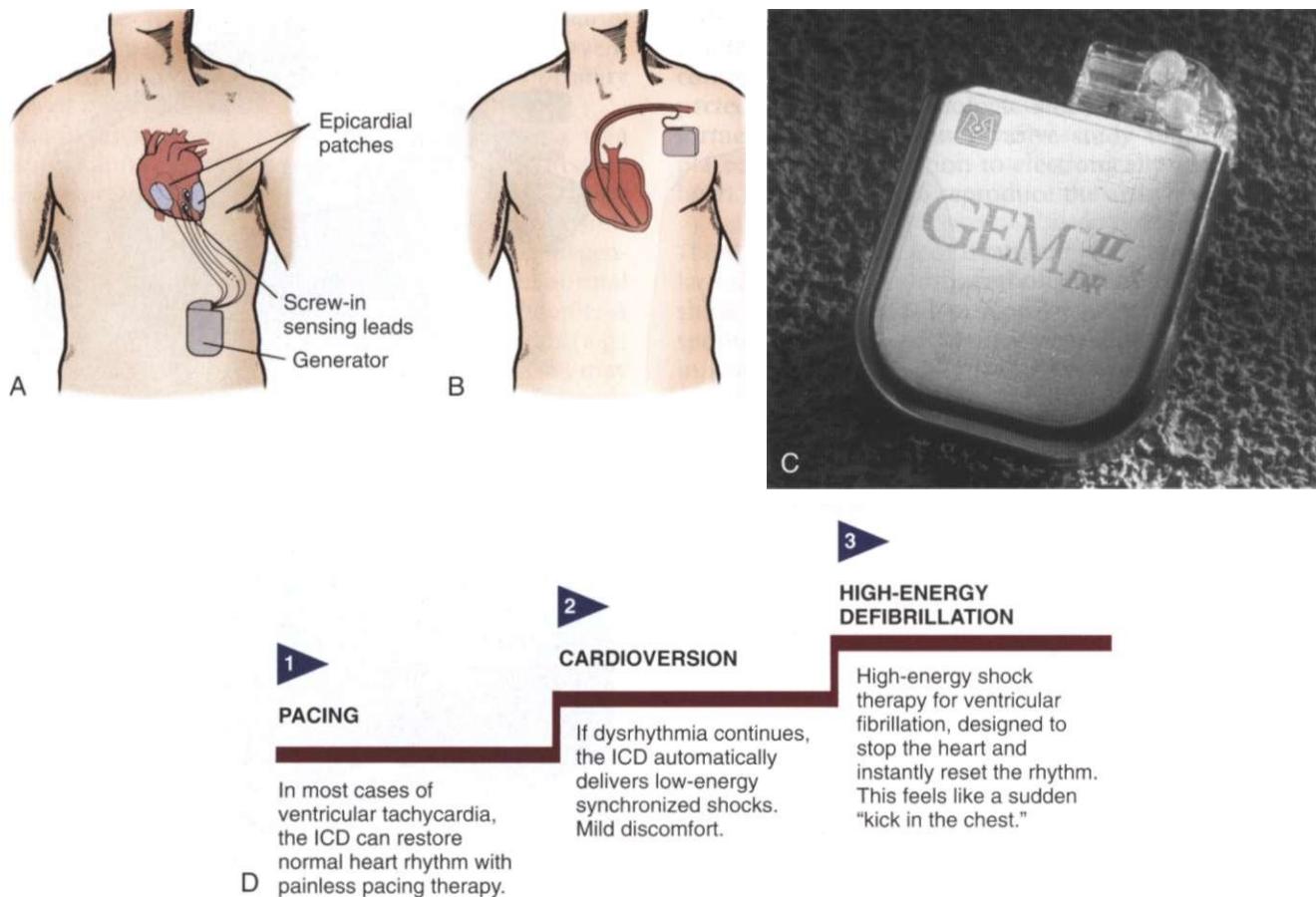


Figure 12-20

A, Placement of an implantable cardioverter defibrillator (ICD) and epicardial lead system. The generator is placed in a subcutaneous "pocket" created in the left upper abdominal quadrant. The epicardial screw-in sensing leads monitor the heart rhythm and connect to the generator. If a life-threatening dysrhythmia is sensed, the generator can pace-terminate the dysrhythmia or deliver electrical cardioversion or defibrillation through the epicardial patches. With this system, the leads/patches must be placed during open chest surgery. **B**, Transvenous lead system. Open chest surgery is not needed to place this unit. The pacing/cardioversion/defibrillation functions are all contained in a lead (or leads) inserted into the right atrium and ventricle. New generators are small enough to place in the pectoral region. **C**, An example of a dual-chamber ICD (Medtronic Gem II DR) with tiered therapy and pacing capabilities. **D**, Tiered therapy is designed to use increasing levels of intensity to terminate ventricular dysrhythmias. (From Urden LD: *Thelan's critical care nursing: diagnosis and management*, ed 5, St Louis, 2006, Mosby. Courtesy Medtronic Inc., Minneapolis, MN.)

estimated 5% of sudden cardiac arrest victims in the United States survive without this treatment. Early defibrillation is the key to survival, and toward that end, emergency medical teams are using portable automatic external defibrillator (AED) units that use a computer program to sense whether a defibrillatory shock is warranted and will initiate the shock.²⁷⁰

The most appropriate and effective drug or drug combination remains unknown, and side effects of long-term rate and rhythm control intervention (e.g., organ toxicity of the lung, liver, and thyroid; aggravation of a preexisting arrhythmia or development of a new arrhythmia instead of preventing it) may prevent long-term use of drug therapy. About 10% of affected individuals continue to have episodes despite treatment, and one half who are treated have a recurrence within 6 months.

2 CARDIOVERSION

1 PACING

In most cases of ventricular tachycardia, the ICD can restore normal heart rhythm with painless pacing therapy.

3 HIGH-ENERGY DEFIBRILLATION

If dysrhythmia continues, the ICD automatically delivers low-energy synchronized shocks. Mild discomfort.

High-energy shock therapy for ventricular fibrillation, designed to stop the heart and instantly reset the rhythm. This feels like a sudden "kick in the chest."

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-12

Arrhythmias

PREFERRED PRACTICE PATTERNS

5A: Primary Prevention/Risk Reduction for Loss of Balance and Falling

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

Any time a person's pulse is abnormally slow, rapid, or irregular, especially in the presence of known cardiac involvement, documentation and notification of the physician are necessary. Early detection and treatment of AF can be critical in reducing the client's risk of stroke and hemodynamic compromise.

Predisposing factors for arrhythmias include fluid and electrolyte imbalance (see Chapter 5) and drug toxicity (see Table 12-5 and Special Implications for the Therapist: Cardiomyopathy). To prevent postoperative cardiac arrhythmias, consult carefully with respiratory therapy personnel to provide adequate oxygen during activities that increase the heart's workload.

Individuals experiencing exercise intolerance due to palpitations, fatigue, and shortness of breath should be assessed further. Keep in mind that people with arrhythmias can be completely asymptomatic. And it is possible that clients describing palpitations or similar phenomena may not be experiencing symptoms of arrhythmic heart disease at all.

Palpitations can occur as a result of an overactive thyroid, secondary to caffeine sensitivity, as a side effect of some medications, from decreased estrogen levels, and through the use of drugs such as cocaine. Encourage the client to report any such symptoms to the physician if this has not already been brought to the physician's attention. See Special Implications for the Therapist: Signs and Symptoms of Cardiovascular Disease.

It is the position of the American Physical Therapy Association that properly trained physical therapists should be authorized to perform advanced cardiac life support procedures, including cardiac monitoring for arrhythmia recognition and cardiac defibrillation.¹⁸

Physical therapists exercise many people with a history of personal or family heart disease or known risk factors for cardiac disease potentially necessitating cardiopulmonary resuscitation. Make sure to have advanced cardiac life support equipment available in case of emergency.

Public access to emergency defibrillation was signed into law (Cardiac Arrest Survival Act; HR 2498) in 2000 placing AEDs in federal buildings and providing nationwide Good Samaritan protection that exempts from liability anyone who renders emergency treatment with a defibrillator to save a life. Also signed into law was the Rural Access to Emergency Devices Act (SF 2528) authorizing \$25 million in federal funds to help rural communities purchase AEDs and train people to use them. Clinics offering physical and occupational therapy services should have an AED on site at all times.²⁷⁰

Performing an assessment of falls for individuals with cardiac disease, especially for anyone with a personal or family history of arrhythmias, is highly recommended. Screening for syncope, assessing balance and fall risk, and falling prevention programs are important components of a therapist's evaluation. If the individual is also on anticoagulation therapy, he or she should be monitored (and taught how to self-monitor) for signs and symptoms of bleeding; in the acute care setting, the therapist can monitor international normalized ratio (INR). Evaluations of specific assessment and screening tests are available.^{72,104,308,338}

Exercise and Arrhythmias

Exercise often increases arrhythmias because of the increase in activity of the sympathetic nervous system and the increase in circulating catecholamines. Exercise may induce cardiac arrhythmias under several specific conditions, including diuretic and digitalis therapy, or following recent ingestion of caffeine.

Exercise-induced arrhythmias are generated by enhanced sympathetic tone, increased myocardial oxygen demand, or both. The therapist can be involved in preparticipation screening of all athletes for conditions that put them at risk for sudden cardiac death (see Special Implications for the Therapist: Cardiomyopathy). At times, the arrhythmias may disappear with exercise and increased perfusion. Exercise recommendations for athletes with selected arrhythmias are available.⁹⁴

Medications that are effective in controlling arrhythmias at rest may not be effective during exertion or stress. In addition, side effects of antiarrhythmic agents may be more apparent during exercise. For example, decreases in either exercise performance or blood pressure during exercise may occur. Because of their effects on the electrophysiologic characteristics of cardiac cells, these medications have the potential to cause abnormal rhythms. The effect of slowing the impulse through the myocardium may manifest itself during exercise as a partial or complete heart block.

Individuals with known arrhythmias and clients who are taking antiarrhythmic medications may need to be evaluated under conditions of graded exercise to ensure that the arrhythmia remains under control during activity. Monitoring heart rate and blood pressure during activity and palpation of peripheral pulses are essential in the absence of ECG.

Continued monitoring and observation during the recovery period are also important, because arrhythmias often occur during recovery rather than during peak exercise. If the exercise is stopped abruptly and the individual remains upright, pooling of blood in the lower body occurs. The decreased venous return and subsequent decreased blood flow to the heart may facilitate an irregular rhythm. By continuing to exercise at a low intensity during recovery, a sudden decrease in venous return is avoided.

For the client who is wearing or has worn a cardiac monitor (Holter, event, loop), the therapist must obtain the interpretation of the results to determine if modifications are needed in the person's activities. Anyone with life-threatening arrhythmias should not begin physical therapy activity until intervention for the arrhythmia is initiated and the condition is stabilized. Increasing frequency of arrhythmias developing with activity must be evaluated by the physician.¹⁴⁹

Pacemaker

For the client wearing a pacemaker, the first weeks after surgery may be characterized by fatigue, during which time activity restrictions apply. Most people can drive, but strenuous activities using the arms (e.g., house-

Continued.

work, golf, tennis, lifting more than 10 lb) are contraindicated. Once the incision is fully healed and the pacemaker is stable, scar mobilization is permissible. The usual precautions for scar mobilization apply, including mobilizing the tissue in the direction of the scar before using any cross-transverse techniques and mobilizing toward the scar rather than away from the scar to avoid overstressing the healing tissue.

Problems with pacemakers are uncommon, but any unusual deviation from the set heartbeat expected or the development of unusual symptoms, such as dyspnea, dizziness or light-headedness, and syncope or near syncope, must be reported immediately to the physician. It is important that the therapist understand the underlying problem as well as the type of pacemaker the client is using before monitoring the client's response to an exercise program. More detailed information regarding types of pacemakers and pacemaker implantation is available¹⁵⁰; see also information from pacemaker manufacturers.

It should be noted that MRIs and prolonged exposure to electromagnetic waves are contraindicated in anyone who is pacemaker dependent. Most exposures to electromagnetic interference are transient and pose no threat to people with pacemakers and implantable cardioverter-defibrillators. Concerns that cellular telephone radiation is linked to pacemaker or implantable cardioverter-defibrillator disruption have not been substantiated or proven clinically important.

Heart rate is limited to the programmed level, and individuals with fixed-rate ventricular synchronous devices require monitoring by blood pressure and perceived exertion scales, with close attention to symptoms of cerebral ischemia. Newer, improved pacemakers produce the cardiac output needed for exercise, making it possible for individuals with pacemakers to be physically active at work and during recreation. Exercise may be limited only by the underlying heart disease and left ventricular function. If the pacemaker recipient has undergone exercise testing safely, aerobic conditioning and endurance training can be initiated, although precaution is still advised regarding vigorous upper-body activities.

In some individuals who have suffered cardiac arrest and now have a pacemaker (or other implantable device), the response to surviving cardiac arrest has been compared to posttraumatic stress disorder (see discussion in Chapter 3) that can occur after a person experiences a traumatic event that is outside the realm of usual human experience. Depression, anxiety, difficulty concentrating, negative health beliefs, and increased somatic complaints may be present with or without persistent emotional disability and maladaptation to the event. The therapist should refer anyone suspected of having persistent depression or anxiety to the physician or mental health professional.

DISEASES AFFECTING THE HEART VALVES

Heart problems that occur secondary to impairment of valves may be caused by infections such as endocarditis, congenital deformity, or disease (e.g., rheumatic fever, coronary thrombosis). Valve deformities are classified as functional (e.g., stenosis, insufficiency) or anatomic (e.g., prolapse; congenital deformities; deformities caused by rheumatic fever, trauma, infection, ischemia) (Fig. 12-21).

Stenosis is a narrowing or constriction that prevents the valve from opening fully and may be caused by scars or abnormal deposits on the leaflets. Valvular stenosis causes obstruction to blood flow, and the chamber behind the narrow valve must produce extra work to sustain cardiac output.

Insufficiency (also referred to as *regurgitation*) occurs when the valve does not close properly and causes blood to flow back into the heart chamber. The heart gradually dilates in response to the increased volume of work; severe degrees of incompetence are possible in the absence of symptoms. *Prolapse* affects the mitral or tricuspid valve and occurs when enlarged leaflets bulge backward into the atrium.

Valve conditions increase the workload of the heart and require the heart to pump harder to force blood through a stenosed valve or to maintain adequate flow if blood is seeping back. Initially the cardiovascular system compensates for the overload and the person remains asymptomatic, but eventually as stenosis or insufficiency progresses, cardiac muscle dysfunction and accompanying symptoms of heart failure (breathlessness, dyspnea) develop.

Over the past 15 years, advances in surgical techniques and a better understanding of timing for surgical intervention have brought tremendous improvement in the clinical outcome of people with valvular heart disease, extending survival rates with less overall morbidity.⁶³

The presence of CAD in clients with either mitral or aortic valve disease is a negative prognostic indicator; ischemic mitral regurgitation carries the worst prognosis, with higher operative mortality and lower long-term survival compared with nonischemic cases.⁶³

Heart transplantation may be necessary when the risk of surgery is prohibitively high in some cases of valvular disease. Continued advances in noninvasive assessment (e.g., RT-3D echocardiography) and noninvasive treatment (e.g., gene therapy, valves grown from blood vessel cells, and even valve self-repair with tissue-engineering techniques) should improve the outlook for anyone with valvular heart disease in the years to come.

Mitral Stenosis

Etiologic Factors and Pathogenesis

Mitral stenosis is a sequela of rheumatic heart disease that primarily affects women. Often a history of rheumatic fever is absent. Because the mitral valve is thickened, it

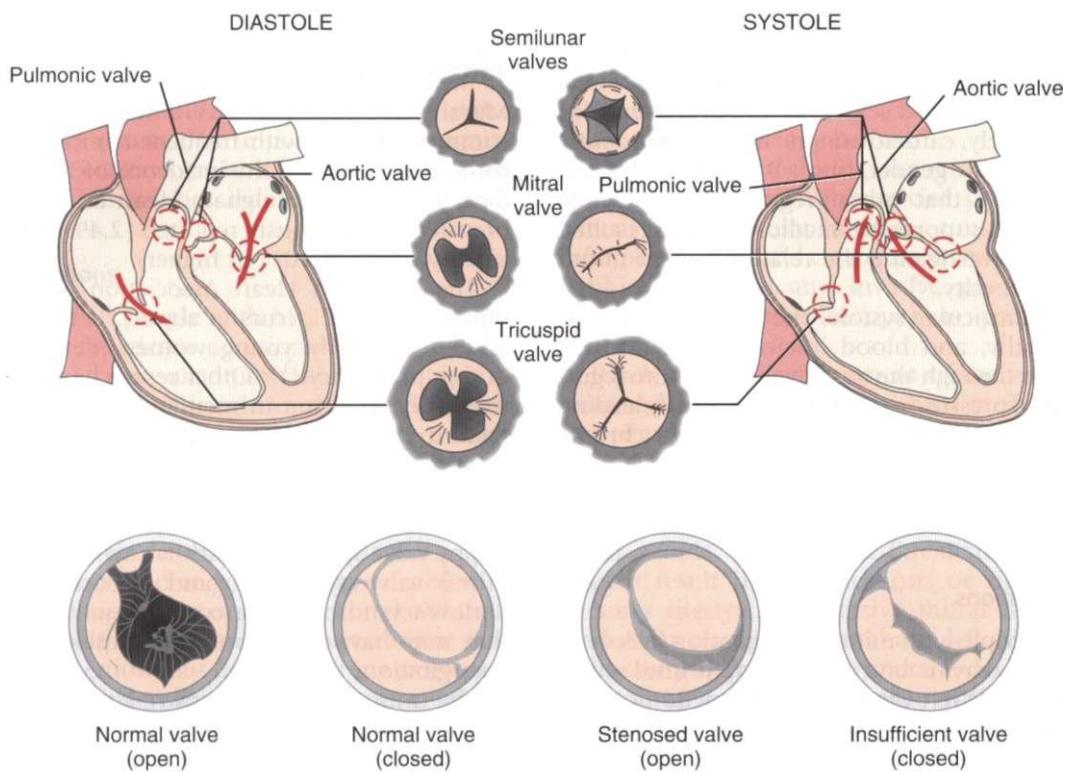


Figure 12-21

Valves of the heart. **A**, The pulmonic, aortic, mitral, and tricuspid valves are shown here as they appear during diastole (ventricular filling) and systole (ventricular contraction). **B**, Normal position of the valve leaflets, or cusps, when the valve is open and closed; fully open position of a stenosed valve; closed regurgitant valve showing abnormal opening into which blood can flow back.

opens in early diastole with a snap that is audible on auscultation and then closes slowly with a resultant murmur.

The anterior and posterior leaflets are fixed like a funnel with an opening in the center, and they move together, rather than in opposite directions. When the valve has narrowed sufficiently, left atrial pressure rises to maintain normal flow across the valve and to maintain a normal cardiac output. This results in a pressure difference between the left atrium and the left ventricle during diastole.

Clinical Manifestations

In mild cases, left atrial pressure and cardiac output remain normal, and the person is asymptomatic, perhaps until pregnancy or the development of AF, when dyspnea and orthopnea develop. In moderate stenosis, dyspnea and fatigue appear as the left atrial pressure rises and mechanical obstruction of filling of the left ventricle reduces cardiac output.

With severe stenosis, left atrial pressure is high enough to produce pulmonary venous congestion at rest and reduce cardiac output, with resulting dyspnea, fatigue, and right ventricular failure. Lying down at night further increases the pulmonary blood volume, causing orthopnea and paroxysmal nocturnal dyspnea.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Echocardiography is the most valuable technique for assessing mitral

valve stenosis and providing information about the condition of the valve and left atrial size. Doppler techniques (measuring blood flow using ultrasound) can be used to determine the severity of the problem.

Because mitral stenosis may be asymptomatic, intervention is delayed until symptoms develop. Mitral stenosis may be present for a lifetime with few or no symptoms, or it may become severe in a few years. The onset of AF accompanied by more severe symptoms may be treated pharmacologically (digoxin, antiarrhythmic agents, anticoagulants).

Surgery may be indicated in the presence of uncontrollable pulmonary edema, severe dyspnea limiting function, pulmonary hypertension, arrhythmia, or systemic emboli uncontrolled by anticoagulation treatment. Surgical procedures include valve repair (commissurotomy to break apart the adherent leaves), replacement with an artificial valve, or balloon valvotomy.

In many cases, balloon valvotomy provides excellent mechanical relief with prolonged benefit, in contrast to the poor results in aortic stenosis.⁶³ Operative mortality rates are low; problems associated with prosthetic valves may occur because of thrombosis, leaking, endocarditis, or degenerative changes in tissue valves.

Mitral Regurgitation

Etiologic Factors and Pathogenesis

Mitral regurgitation has many possible causes, but involvement of the mitral valve from ischemic heart

disease accounts for approximately one half of all cases. Other secondary causes include infective endocarditis (valve perforation), dilated cardiomyopathy, rheumatic disease, collagen vascular disease, rupture of the chordae tendineae, and, rarely, cardiac tumors. It is independently associated with female gender, lower BMI, and older age. Evidence suggesting that mitral regurgitation may be induced by appetite-suppressant medications has resulted in new research investigating the relationship of mitral regurgitation to obesity.¹⁶⁸

During left ventricular systole, the mitral leaflets do not close normally, and blood is ejected into the left atrium as well as through the aortic valve. In acute regurgitation, left atrial pressure rises abruptly, possibly leading to pulmonary edema. When regurgitation is a chronic condition, the left atrium enlarges progressively; the degree of enlargement usually reflects the severity of regurgitation.

Clinical Manifestations

Unfortunately, people with mitral regurgitation lack early warning signs and may remain asymptomatic until severe and often irreversible left ventricular dysfunction occurs. For many years the left ventricular end-diastolic pressure and the cardiac output may be normal at rest, even with considerable increase in left ventricular volume. Eventually, left ventricular overload may lead to left ventricular failure. People with mitral regurgitation experience exertional dyspnea (because of increased left atrial pressure) and exercise-induced fatigue (because of reduced cardiac output). AF may also develop.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. The diagnosis is primarily clinical (auscultation), but it can be confirmed and quantified by color Doppler echocardiography. Other testing procedures may include cardiac catheterization to assess the regurgitation, left ventricular function, and pulmonary artery pressure; coronary arteriography to determine the cause of the lesion and for preoperative evaluation; and nuclear medicine techniques to measure left ventricular function and estimate the severity of regurgitation.

Persons with chronic lesions who are asymptomatic require careful monitoring for left ventricular function and may require surgery even if no symptoms are present. Unlike stenosis, regurgitant lesions may progress insidiously, causing left ventricular damage before symptoms have developed.

Surgical intervention may be recommended if left ventricular function is impaired or when activity becomes severely limited. Mitral valve repair has a lower operative mortality and a better late outcome than mitral valve replacement. Acute mitral regurgitation secondary to MI often requires emergency surgery, but the surgical risk is high and the outcome poor. Acute non-MI-related mitral regurgitation has a much better prognosis with higher postoperative survival after well-timed mitral valve repair. Indicators of poorer prognosis include mitral valve replacement, age more than 75 years, and the presence of CAD.⁶³

Mitral Valve Prolapse

Incidence and Etiologic Factors

Mitral valve prolapse (MVP) has been described as a common disease with frequent complications. There is some dispute about the incidence of MVP. According to data from the Framingham Heart Study, MVP is not as prevalent as previously reported (2.4% compared to previously reported 10% or higher).

The American Heart Association and other sources report that MVP occurs in about 2% to 6% of "normal" adults, especially young women, detected most often during pregnancy.^{13,65} Other researchers report that MVP is equally common in men and women, although men seem to have a higher incidence of complications.¹⁴⁴

MVP is characterized by a slight variation in the shape or structure of the mitral (left atrioventricular) valve. This structural variation has many other names, including floppy valve syndrome, Barlow's syndrome, myxomatous mitral valve syndrome, and click-murmur syndrome. Barlow's syndrome is a controversial clinical syndrome that may have as its only manifestation MVP without regurgitation.

The cause remains unknown, although there may be a genetic component involving the angiotensin receptor gene resulting in autonomic or neuroendocrine dysfunction.³²³ Results of family studies of people with MVP favor an autosomal dominant pattern of transmission for primary MVP with nearly 100% gene expression by females.⁹⁷ This condition usually occurs in isolation; however, it can be associated with a number of other conditions, such as Marfan's syndrome, rheumatic fever, endocarditis, myocarditis, atherosclerosis, SLE, muscular dystrophy, acromegaly, adult polycystic kidney disease, and cardiac sarcoidosis.

Pathogenesis

MVP is a pathologic, anatomic, and physiologic abnormality of the mitral valve apparatus affecting mitral valve leaflet motion. Normally, when the lower part of the heart contracts, the mitral valve remains firm and prevents blood from leaking back into the upper chambers. In MVP, the slight variation in shape of the mitral valve allows one part of the valve, the leaflet, to billow back into the left atrium during contraction of the ventricle. One or both of the valve leaflets may bulge into the left atrium during ventricular systole. Usually the amount of blood that leaks back into the left atrium is not significant, but in a small number of people, it develops into mitral regurgitation. MVP is the most common cause of isolated mitral regurgitation.

The presence of symptoms linked to neuroendocrine dysfunctions or to the autonomic nervous system has led to the recognition of a pathologic condition known as *mitral valve prolapse syndrome (MVPS)*. Usually diagnosed by chance in asymptomatic individuals during routine tests, MVPS (prolapse with or without mitral regurgitation) has a high clinical incidence of neuropsychiatric symptoms (e.g., anxiety disorder, panic attacks, depression), as well as symptoms of autonomic dysfunction (e.g., postural hypotension, palpitations, cold hands and feet, shortness of breath, chest pain).

As the autonomic nervous system is being formed in utero, the mitral valve is also being formed. If there is a slight variation in the structure of the heart valve, there is also a slight variation in the function or balance of the autonomic nervous system. The importance of recognizing that MVP may occur as an isolated disorder or with other coincident findings has led to the use of both terms.

Clinical Manifestations

More than 50% of all people with MVP are asymptomatic, another 40% experience occasional symptoms that are mildly to moderately uncomfortable, and only 1% suffer severe symptoms and lifestyle restrictions. Although the malformation occurs during gestation, it usually remains unnoticed until young adulthood. The person usually becomes aware of symptoms suddenly, and there does not appear to be any correlation between the severity of symptoms and the severity of the prolapse.

The most common triad of symptoms associated with MVP is profound fatigue that cannot be correlated with exercise or stress, palpitations, and dyspnea. Fatigue may not be related to exertion, but deconditioning from prolonged inactivity may develop, further complicating the picture.

The therapist is more likely to see the individual with MVP associated with connective tissue disorders or the MVPS with autonomic dysfunction. Frequently occurring musculoskeletal findings in clients with MVPS include joint hypermobility, temporomandibular joint (TMJ) syndrome, pectus excavatum, mild scoliosis, straight thoracic spine, and myalgias. The increased joint mobility that has been identified in a small proportion of persons with MVP does not appear to lead to either severe arthritis or frequent joint dislocations.⁹⁸

Other symptoms associated with MVPS may include tremors, swelling of the extremities, sleep disturbances, low back pain, irritable bowel syndrome, excessive perspiration or inability to perspire, rashes, muscular fasciculations, visual changes or disturbances, difficulty in concentrating, memory lapses, and dizziness.

Chest pain or discomfort may occur as a result of autonomic nervous system dysfunction (dysautonomia). The autonomic nervous system imbalance results in inadequate relaxation between respirations and eventually causes the chest wall muscles to go into spasm. The chest pain is sharp, lasts several seconds, and is usually felt to the left of the sternum. It is intermittent pain that may occur frequently for a few weeks and then disappear completely, only to return again some weeks later.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. MVP is often discovered during routine cardiac auscultation or when echocardiography is performed for another reason. It is characterized by a symptomatic clinical presentation and clicking noise on auscultation in late systole, with or without sounds of valvular leak (murmur).

The mitral valve begins to prolapse when the reduction of left ventricular volume during systole reaches a critical point at which the valve leaflets no longer coapt (edges approximate together); at that instant, the click occurs

and the murmur begins. Complete diagnostic (major and minor) criteria have been outlined elsewhere.⁴⁸ Echocardiography may be used to confirm the diagnosis, and ECG, event, or Holter monitoring (see Fig. 12-19) to show arrhythmias may be used.

Management includes reassurance; β -blockers to control arrhythmias; an exercise program to improve overall cardiovascular function; counseling to eliminate caffeine, alcohol, and cigarette use; and administration of antibiotics before any invasive procedure (including dental work, sigmoidoscopy) as prophylaxis against endocarditis.

Rarely, surgical replacement of the valve may be recommended when severe structural problems are present that contribute to reduced activity or deterioration of left ventricular function from progression of MVP to mitral regurgitation.

MVP or MVPS is a benign condition in the vast majority of people. It is not life-threatening and only rarely does it result in complications or significantly alter a person's lifestyle. Progressive mitral regurgitation with gradual increase in left atrial and left ventricular size, AF, pulmonary hypertension, and the development of CHF occur in 10% to 15% of people with both murmurs and clicks. Men older than 50 years are most often affected.⁴⁸

According to new data available, people with MVP or MVPS are not at greater risk for heart failure, other forms of heart disease, or early death from stroke as was once thought.

Aortic Stenosis

Etiologic Factors and Pathogenesis

Aortic stenosis is a disease of aging that is likely to become more prevalent as the proportion of older people in our population increases. It is most commonly caused by progressive valvular calcification either superimposed on a congenitally bicuspid valve or, in the older adult, involving a previously normal valve following rheumatic fever.

Other risk factors for aortic stenosis are the same as those for heart disease and include obesity, a sedentary lifestyle, smoking, and high cholesterol. Factors affecting the progression of the disease remain uncertain. Over 80% of affected persons are men, and when women are affected, differences are noted (e.g., women with aortic stenosis have thicker ventricular walls reducing wall stress and higher ejection fractions) that require different post-operative management (e.g., low cardiac output requiring volume expansion rather than the use of pressor agents).⁶³ Ejection fraction is the amount of blood the ventricle ejects; the normal ejection fraction is about 60% to 75%. A decreased ejection fraction is a hallmark finding of ventricular failure.

Although the deformed valve is not stenotic at birth, it is subjected to abnormal hemodynamic stress, which may lead to thickening and calcification of the leaflets with reduced mobility. The orifice of the aortic valve narrows, causing increased resistance to blood flow from the left ventricle into the aorta.

Outflow obstruction increases pressure within the left ventricle as it tries to eject blood through the narrow opening, causing decreased cardiac output, left ventricular hypertrophy, and pulmonary vascular congestion. Preschool and school-aged children are more likely to have a bicuspid valve; teenagers and young adults present with three leaflets, but these are partially fused.

Clinical Manifestations

In adults, aortic stenosis is usually asymptomatic until the sixth (or later) decade. Characteristic sounds may be heard on auscultation, but cardiac output is maintained until the stenosis is severe and left ventricular failure, angina pectoris, or exertional syncope develops. The origin of exertional syncope in aortic stenosis remains controversial; it is perhaps caused by an exercise-induced decrease in total peripheral resistance, which is uncompensated because cardiac output is restricted by the stenotic valve. The most common sign of aortic stenosis is a systolic ejection murmur radiating to the neck (usually heard best in the aortic area). Sudden death may occur, even in previously asymptomatic individuals.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. The clinical assessment of aortic stenosis can be difficult, especially in the older person. Echo Doppler (echocardiography with Doppler ultrasonography) is diagnostic in most cases. ECG may show left ventricular hypertrophy, and x-ray or fluoroscopy may show a calcified aortic valve. Coronary angiography may be necessary in older adults at risk for coronary disease before valve replacement.

Pharmacologic therapy has limited use in this condition. Surgical intervention is usually required for the symptomatic person and should be strongly considered for the asymptomatic person because of the risk of sudden death.

Surgical procedures may include valve replacement with a mechanical prosthesis or bioprosthetic (made with biologic material) or use of the pulmonary valve in place of the aortic valve and replacement of the pulmonary valve with a homograft (Ross procedure). Homografts have been shown to have a superior durability compared to xenogenic biologic prostheses. Approximately 40% were still in place 20 years after implantation in the aortic position. Their low rate of thromboembolic events made life-time anticoagulative therapy unnecessary, and their hemodynamics may be superior to that of all other heart valve prostheses.¹³⁷

A less invasive alternative to aortic valve replacement, balloon valvuloplasty (splitting the stenotic valve with a balloon-tipped catheter that is introduced into the valve and inflated), has been used in treatment for aortic stenosis. Limited long-term success with less reliable and less durable outcomes has resulted in this procedure's being considered palliative rather than curative. It may be useful in alleviating symptoms in people who are not candidates for aortic valve replacement because of other medical problems.

Adults with aortic stenosis who are asymptomatic have a normal life expectancy; they should receive prophylactic antibiotics against infective endocarditis. Once symp-

toms appear, the prognosis is poor without surgery but excellent with valve replacement even in the older adult, especially in the absence of coexisting illnesses.⁶³

The onset of angina, exercise-induced syncope, or cardiac failure indicates a poor prognostic outcome resulting in death. Mortality rises to 10% after age 80 years. Bioprostheses may develop degenerative changes, requiring replacement in 2 to 20 years. This is quite variable and depends on the person's age at the time of implantation.

Aortic Regurgitation (Insufficiency)

Etiologic Factors and Pathogenesis

In the past, aortic regurgitation occurred secondary to rheumatic fever, but antibiotics have reduced the number of rheumatic fever-related cases. Nonrheumatic causes account for most cases today, including congenitally bicuspid valves, infective endocarditis (valve destruction by bacteria), and hypertension. Aortic regurgitation may also occur secondary to aortic dissection with or without aortic aneurysm (see Fig. 12-27), ankylosing spondylitis, Reiter's syndrome, collagen vascular disease, syphilis, and Marfan's syndrome.

When cardiac systole ends, the aortic valve should completely prevent the flow of aortic blood back into the left ventricle. A leakage during diastole is referred to as *aortic regurgitation* or *aortic insufficiency*. When aortic regurgitation develops gradually, the left ventricle compensates by both dilation and enough hypertrophy to maintain a normal wall thickness/cavity ratio, thereby preventing development of symptoms. Eventually the left ventricle fails to stand up under the chronic overload, and symptoms develop.

Clinical Manifestations

Longstanding aortic regurgitation may remain asymptomatic even as the deformity increases, causing enlargement of the left ventricle. The large total stroke volume in aortic regurgitation produces a wide pulse pressure and systolic hypertension, resulting in exertional dyspnea, fatigue, and excessive perspiration with exercise as the most frequent symptoms; paroxysmal nocturnal dyspnea and pulmonary edema may also occur. Angina pectoris or atypical chest pain may be present, but this is uncommon in the absence of CAD.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Once aortic regurgitation is suspected on physical examination, echocardiography with Doppler examination of the aortic valve can help estimate its severity. Aortography during catheterization helps confirm the severity of the disease. Scintigraphic studies can quantify left ventricular function and functional reserve during exercise and provide a useful predictor of prognosis.

Acute aortic regurgitation may lead to left ventricular failure; surgical reconstruction or replacement of the valve (Ross procedure; see the section on Aortic Stenosis) is advisable before onset of permanent left ventricular damage (usually before ejection fraction falls below 55%), even in asymptomatic cases. Chronic regurgitation

carries a poor prognosis without surgery when significant symptoms develop. Medical therapy may include vasodilators to reduce the severity of regurgitation and diuretics and digoxin to stabilize or improve symptoms.

Tricuspid Stenosis and Regurgitation

Tricuspid stenosis may be congenital or rheumatic in origin and is uncommon. Exercise testing and rehabilitation do not occur until after valve surgery. Tricuspid regurgitation may occur secondary to carcinoid syndrome, SLE, or infective endocarditis among injection drug users, and in the presence of mitral valve disease. Surgical repair is more common than valvular replacement for tricuspid valve disease.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-13

Valvular Heart Disease

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6D: Impaired Aerobic Capacity/Endurance Associated with Cardiovascular Pump Dysfunction or Failure

People with mild valvular malfunction have no symptoms and can usually exercise vigorously and take part in intense sports activities without adverse effects. Although exercise will not improve the mechanical function of a valve, improvement in submaximal cardiac capacity can occur. Exercise is usually stopped for the same reason as it is in healthy adults (i.e., when respiratory distress is obvious or when the person expresses a desire to stop).

Involvement of more than one valve is not uncommon in people with rheumatic valvular disease and in people who develop valvular regurgitation as a result of ventricular dilation. Usually symptoms and clinical course are determined by the predominant pathologic condition. When two valves are affected equally, symptoms are determined by the most proximally located valve. The combination of aortic regurgitation with mitral valve regurgitation is the most common, but the combination of mitral valve disease (either regurgitation or stenosis) with aortic stenosis is the most problematic.³⁰³

Exercise

Exercise testing for most people with valvular disease is of limited value. For example, there is poor correlation between the degree of mitral stenosis and the duration of symptom-limited treadmill exercise. However, exercise echocardiography performed while the individual is on a stationary cycle can be a valuable means for determining left ventricular function in people during exercise.

Prescriptive exercise must be individualized based on the underlying pathologic condition, medical intervention, and condition of the person. General guidelines include exercise a minimum of 3 days/wk with alternate days of rest to allow for maximum recuperation. Walking, biking, and swimming are acceptable

exercise modalities, but weight training may be considered contraindicated in anyone who is symptomatic with shortness of breath or chest pain or discomfort.

A perceived exertion between light and somewhat hard (RPE of 11 to 14; see Table 12-13) is the goal, but the individual will usually begin with a much lighter workout and progress over time to this level.³⁰³ Tolerance to symptoms and current exercise habits are important determinants in progressing an exercise program.

Some people with valvular disease avoid physical activity as much as possible and never exercise to the point of developing any symptoms of dyspnea, fatigue, or muscular discomfort. These symptoms develop at light loads in people unaccustomed to any physical activity, regardless of the severity of the valvular disease. Other people force themselves to ignore mild (or even moderate to severe) symptoms to stay on the job or finish a task started.

Fatigue, weakness, and pallor are signs of an inadequate cardiac output for the demands of the exercise. These signs and symptoms are partly subjective, and it is a clinical decision as to how far to allow these people to continue exercising. Chest pain may indicate myocardial ischemia or pulmonary hypertension, or it may be a noncardiac symptom arising from the chest wall.

Follow precautions for angina pectoris. Exercise should be stopped immediately when any signs of reduced cerebral blood flow develop, such as severe facial pallor, confusion, dizziness, heart palpitations, or unsteady gait (see also Box 12-4 and Box 12-9).

Pulmonary edema can be produced by exercising beyond a certain point in people with valvular disease, especially those with mitral stenosis. Pulmonary congestion induced by exercise may cause coughing rather than dyspnea, and exercise should be stopped if coughing becomes significant. Heart failure may occur secondary to chronic, progressive valvular disease. Slight puffiness of the ankles at the end of the day, nocturia, mild nocturnal dyspnea, unexpected weight gain, or more than the usual amount of fatigue can be minor symptoms that are passed over unless specifically sought. Such symptoms must be reported to the physician. (See also Special Implications for the Therapist: Congestive Heart Failure.)

The status of the myocardium is another important variable in exercise impairment relative to valvular heart disease. Severe aortic regurgitation is well tolerated for many years until myocardial weakness occurs. In all forms of heart disease, the healthy myocardium can compensate and maintain the systemic blood flow at or near normal levels for an extended period of time. For the client with valvular disease and myocardial disease or associated CAD, this compensation is not possible, and a lower exercise capacity results.

Stenosis

Valvular stenosis develops or progresses gradually, and because the normal valve orifice is larger than is neces-

Continued.

sary, stenosis is usually severe before exercise symptoms occur (i.e., a normal valve is larger than is needed for normal functioning and therefore has excess capacity). Stenosis only becomes symptomatic when the condition encroaches on the critical cross-sectional diameter of the opening so that a doubling of the blood flow (from activity or exertion) across the valve quadruples the atrial pressure. When the atrial pressure exceeds 25 mm Hg, dyspnea develops. The intensity of exertion associated with dyspnea does correlate with the magnitude of atrial pressure, providing a good indicator of the severity of stenosis. However, some people do not complain of dyspnea from lung congestion, only muscular fatigue on exertion as a result of a low cardiac output.

Stress testing may be performed before initiation of an exercise program; with or without those test results, clients should be monitored closely, possibly using the perceived exertion or dyspnea scales mentioned earlier in this chapter. Because of reduced cardiac output, muscle perfusion is reduced and lactate is produced at low workloads. Maximal heart rate may be reduced when dyspnea is the cause of premature termination of exercise. Exercise systolic blood pressure may reach only 130 mm Hg because of low output. Exercise capacity in clients with mitral stenosis can be improved by slowing heart rate and prolonging the diastolic filling period with the use of β -blocking agents.

In the case of symptomatic aortic stenosis, clients are not candidates for exercise programs because of the danger of sudden death. Persons who are asymptomatic must be carefully screened before increasing their physical activity, and for most, exercise intensity should be mild. In people with impaired left ventricular function, cardiac output fails to increase normally with exercise, causing fatigue. Angina with exercise is a common symptom when the aortic stenosis is severe.

Regurgitation

Exercise capacity may be unaffected in cases of mild regurgitation. Mitral regurgitation increases when aortic blood pressure is increased, such as occurs during isometric contractions. Light to moderate rhythmic and repetitive exercise reduces peripheral resistance and is recommended in place of isotonic exercise, which increases the heart rate. Persons with aortic regurgitation caused by weakening of the aortic wall (Marfan's or Ehlers-Danlos syndrome) must avoid all strenuous exercise.^{95,250}

Prolapse

Most people with MVP can participate in all sports activities, including intense competitive sports. Exercise is a key component in the management of MVP (not to alter function of the prolapsed valve but to improve overall cardiovascular function), and although many clients are referred to an exercise physiologist, the physical therapist may also encounter requests for conditioning and exercise programs. Many times, symptoms of fatigue and dyspnea cause a person to limit physical activity, leading to deconditioning and

contributing to a cycle of even more fatigue and shortness of breath.

Caution is advised in the use of weight training for the client with MVP; gradual buildup using light weights and increased repetitions is recommended. Some people with MVPS are prone to exercise-induced arrhythmias, which can (rarely) result in sudden death. Any time tachycardia develops in someone with known MVP, immediate medical referral is necessary.

Postoperative Considerations

Postoperative considerations are the same as for people who have had abdominal or cardiothoracic surgery (see the section on The Cardiac Client and Surgery in this chapter; see also Box 12-3 and Box 12-4). After uncomplicated valve ballooning, a return to normal activities is possible within 5 to 7 days. Gradual walking programs can be initiated at home for most people 10 days after surgery, or the client may enroll in a structured cardiac rehabilitation program.

Cardiac rehabilitation postoperatively in people with valvular heart disease is similar to that in post-CABG clients. Care should be taken to avoid high-impact exercises or exercises with a risk of trauma in people who are receiving anticoagulation therapy to avoid hemarthrosis and bruising (see Tables 40-8 and 40-9).³⁰³

Exercise outcomes differ after aortic, mitral, and mitral/aortic valve surgery. The degree of improvement in exercise capacity depends on the degree of residual dysfunction, presence or absence of arrhythmia, age of the subject, and the effort made to improve exercise capacity. Functional capacity is substantially increased following aortic valve surgery but limited following mitral and mitral/aortic surgery, possibly because of differences in oxygen uptake. As mentioned, for people with mitral stenosis, exercise provides an early warning system, since the onset of dyspnea with strenuous exercise signals the beginning of clinical deterioration.³⁰³

People with mechanical prosthetic valves receive lifelong anticoagulant therapy (not required for bioprostheses) and may not tolerate vigorous, weight-bearing activities. Mechanical prostheses have fixed openings that place some limitation, at least theoretically, on cardiac performance during maximal effort.¹⁷⁶ Because stress testing results can be normal, exercise Doppler echocardiography has been used to help prescribe physical activity in clients with prosthetic valves.

Infective Endocarditis

Infective, or bacterial, endocarditis is an infection of the endocardium, the lining inside the heart, including the heart valves; it most commonly damages the mitral valve, followed by the aortic, tricuspid, and pulmonic valves. Bacterial endocarditis may involve normal valves but more often affects valves that have been damaged by some other previous pathologic process (e.g., rheumatic disease, congenital defects, cardiac surgery).