

Table 12-16 Clinical Manifestations of Infective Endocarditis

Systemic Infection	Intravascular Involvement	Immunologic Reaction	Musculoskeletal	Neurologic
Fever	Chest pain	Arthralgia	Arthralgia	Confusion
Chills	Congestive heart failure	Proteinuria	Myalgias	Abscess
Sweats	Cold and painful extremities	Hematuria	Low back pain	Cerebritis
Malaise	Clubbing	Acidosis		Meningitis
Weakness	Petechiae	Arthritis		Stroke (embolic or hemorrhagic)
Anorexia				
Weight loss				
Cough				
Dyspnea				
Hemoptysis				

Endocarditis is categorized as either acute or subacute, depending on the clinical course, organisms, and condition of the valves. Endocarditis can occur at any age but rarely occurs in children; one half of all clients diagnosed are older than 60 years. Older adults may be at greater risk of endocarditis because valvular endocardial disruption is more common, immunity is impaired, and nutrition is poor. Endocarditis is more prevalent among men than women.

Etiologic and Risk Factors

Endocarditis is frequently caused by bacteria (particularly streptococci or staphylococci) normally present in the mouth, respiratory system, or GI tract or as a result of abnormal growths on the closure lines of previously damaged valves (e.g., rheumatic disease).

In addition to those with previous valvular damage, persons with prosthetic heart valves, injection drug users, immunocompromised clients (including individuals receiving treatment for cancer), women who have had a suction abortion or pelvic infection related to intrauterine contraceptive devices, and postcardiac surgical clients are at high risk for developing endocarditis. Congenital heart disease and degenerative heart disease, such as calcific aortic stenosis, may also cause endocarditis.

Hospital-acquired infective endocarditis has become more common as a result of iatrogenic endocardial damage produced by surgery, intracardiac pressure-monitoring catheters, ventriculoatrial shunts, and hyperalimentation lines that reach the right atrium. Portals of entry for microorganisms are also provided by wounds, biopsy sites, pacemakers, IV and arterial catheters, indwelling urinary catheters, and intratracheal airways.

Pathogenesis

As an infection, endocarditis causes inflammation of the cardiac endothelium with destruction of the connective tissue. As these bloodborne microorganisms adhere to the endocardial surface, destruction of the connective tissue occurs as a result of the action of bacterial lytic enzymes. The surface endocardium becomes covered with fibrin and platelet thrombi that attract even more thrombogenic material.

The result is the formation of wartlike growths called *vegetations*. These vegetations, consisting of fibrin and

platelets, can break off from the valve, embolize, and cause septic infarction in the myocardium, kidney, brain, spleen, abdomen, or extremities. These thromboemboli contain bacteria that not only cause ischemic infarcts but also form new sites of infection transforming into micro-abscesses. Bacteria may further invade the valves, causing intravalvular inflammation, destroying portions of the valves, and causing valve deformities.

Splinter hemorrhages of the nail beds may be caused by distal vasospasm, embolic events, or other local factors promoting engorgement and bleeding of the capillaries that lie right below the nail. The cause of digital clubbing is unclear, but perhaps platelet clumps lodge in the nail bed capillaries of the fingers and toes and release platelet-derived growth factor, resulting in the pathologic changes of clubbed digits.

Petechiae (small, red, nonblanching macules on the conjunctivae, palate, buccal mucosa, heels, shoulders, arms, legs, and upper chest) are thought to involve microemboli, but some have also suggested that immune complex vasculitis is the primary process.¹⁷³ Infective endocarditis of the right-side heart valves occurs commonly in injection drug users. Although a variety of hypotheses have been put forward to explain this phenomenon, no single explanation has been proven.

Clinical Manifestations

Endocarditis can develop insidiously, with symptoms remaining undetected for months, or it can cause symptoms immediately, as in the case of acute bacterial endocarditis. Clinical manifestations can be divided into many groups (Table 12-16). It causes varying degrees of valvular dysfunction and may be associated with manifestations involving any number of organ systems, including lungs, eyes, kidneys, bones, joints, and CNS. The mitral, aortic, tricuspid, and pulmonic valves can be affected (descending order); more than one valve can be infected at the same time. Neurologic signs and symptoms are predominant in about one third of all cases in those people over 60 years. The classic findings of fever, cardiac murmur, and petechial lesions of the skin, conjunctivae, and oral mucosa are not always present.

Up to 50% of people with infective endocarditis initially have musculoskeletal symptoms, including arthralgia (most common), arthritis, low back pain, and

myalgias. One half of these people will have only musculoskeletal symptoms without other manifestations of endocarditis. The early onset of joint pain and myalgia as the first sign of endocarditis is more likely if the person is older and has had a previously diagnosed heart murmur.

Proximal joints are most often affected, especially the shoulder, followed by knee, hip, wrist, ankle, metatarsophalangeal and metacarpophalangeal joints, and acromioclavicular joints (order of declining incidence). Most often one or two joints are painful, and symptoms begin suddenly, accompanied by warmth, tenderness, and redness. Symmetric arthralgia in the knees or ankles may lead to a diagnosis of rheumatoid arthritis, but as a rule, morning stiffness is not as prevalent in clients with endocarditis as in those with rheumatoid arthritis or polymyalgia rheumatica.

Bone and joint infections are particularly common among injection drug users. The most common sites of osteoarticular infections are the vertebrae, wrist, and sternoclavicular and sacroiliac joints, often with multiple joint involvement.²⁹⁰

Almost one third of clients with endocarditis have low back pain, which may be the primary symptom reported. Back pain is accompanied by decreased range of motion and spinal tenderness. Pain may affect only one side, and it may be limited to the paraspinal muscles. Endocarditis-induced back pain may be very similar to that associated with a herniated lumbar disk, since it radiates to the leg and may be accentuated by raising the leg or by sneezing, coughing, or laughing; however, neurologic deficits are usually absent in persons with endocarditis.

Endocarditis may produce destructive changes in the sacroiliac joint characterized by pain localized over the sacroiliac, probably as a result of seeding of the joint by septic emboli. Widespread diffuse myalgia may occur during periods of fever, but this is not appreciably different from the general myalgia seen in clients with other febrile illnesses. More commonly, myalgia is restricted to the calf or thigh. Bilateral or unilateral leg myalgias occur in approximately 10% to 15% of all persons with endocarditis.

The cause of back pain and leg myalgia associated with endocarditis has not been determined. Concurrent aseptic meningitis is a possible hypothesis; a role for emboli that break off from the infected cardiac valves is supported by biopsy evidence of muscle necrosis or vasculitis in clients with endocarditis. Rarely, other musculoskeletal symptoms, such as osteomyelitis, tendinitis, hypertrophic osteoarthropathy, bone infarcts, and ischemic bone necrosis, may occur.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Infective endocarditis is often difficult to diagnose, since it can present with a wide array of signs and symptoms, as well as a confusing clinical picture. Blood cultures to identify specific pathogens in the presence of septicemia are required to determine appropriate antibiotic therapy, which is the primary medical intervention.

Other laboratory test results indicative of infectious endocarditis include elevated erythrocyte sedimentation

rate, proteinuria, and hematuria. Echocardiography may be used to confirm the diagnosis and is useful in showing underlying valvular lesions and quantifying their severity. This test is not as useful in older adults, because it is common to find echogenic areas around and on degenerative valves that are impossible to distinguish from the infective vegetations seen in infective endocarditis. Large masses on valves are much more diagnostic.

Although it is easily prevented (for the at-risk person) by taking antibiotics before and after procedures such as dental cleaning, genitourinary instrumentation, and open cardiovascular surgery, endocarditis is difficult to treat and can result in serious heart damage or death. Potential complications are many, including CHF and arterial, systemic, or PEs. Therapy with antibiotics may be prolonged, and without complete treatment, relapse can occur up to 2 or more weeks after medical intervention. Surgical valve replacement may be necessary, depending on the response to treatment, sites of infection, recurrent infection, or infection of a prosthetic valve.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-14

Infective Endocarditis

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

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

In the presence of neurologic and/or musculoskeletal manifestations, other practice patterns may be appropriate (see Table 12-16).

Physical exertion beyond normal activities of daily living is usually limited for the person receiving antibiotic therapy for endocarditis and during the following weeks of recovery. The therapist is not likely to treat a person diagnosed during this acute phase of endocarditis. However, because early manifestations of endocarditis may be primarily peripheral (musculoskeletal or cutaneous) in nature, the therapist may be the first to recognize signs and symptoms of a systemic disorder.

Splinter hemorrhages (dark red linear streaks resembling splinters under the nail bed), clubbing (see Fig. 15-4), petechiae, purplish red subcutaneous nodes on the finger and toe pads, and lesions on the thenar and hypothenar eminences of the palms, fingers, and sometimes the soles are present in up to 50% of affected individuals.¹⁷³

For any client with known risk factors or a recent history of endocarditis, the therapist must be alert for signs of endocarditis, indications of complications (easy fatigue associated with heart failure or peripheral emboli), lack of response to therapy intervention, or signs indicating relapse. Often, the client thinks the symptoms are recurrent bouts of the flu.

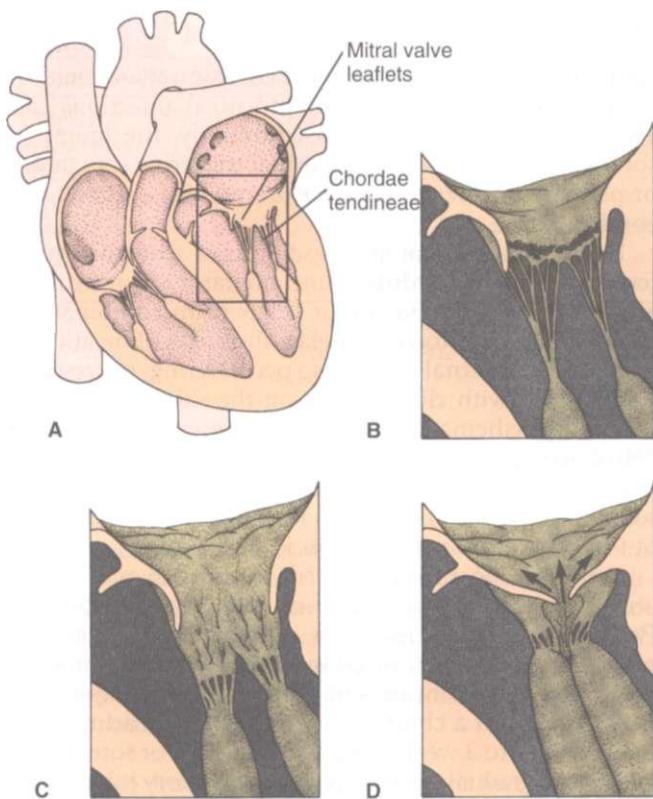


Figure 12-22

Cardiac valvular disease caused by rheumatic fever. **A**, Inflammation of the membrane over the mitral (and aortic) valves may cause edema and accumulation of fibrin and platelets on the chordae tendineae. **B**, This accumulation of inflammatory materials produces rheumatic vegetations that affect the support provided by the chordae tendineae to the atrioventricular valves. **C**, In this view, the mitral valve leaflets have become thickened with scar tissue and calcified. The chordae tendineae often fuse. **D**, As a result, the scarred valve fails to close tightly (mitral stenosis) and regurgitation or backflow of blood into the atrium develops. Prolonged, severe stenosis with mitral regurgitation leads to symptoms of congestive heart failure. (Modified from Goodman CC, Snyder TE: *Differential diagnosis in physical therapy*, ed 3, Philadelphia, 2000, Saunders, p 110.)

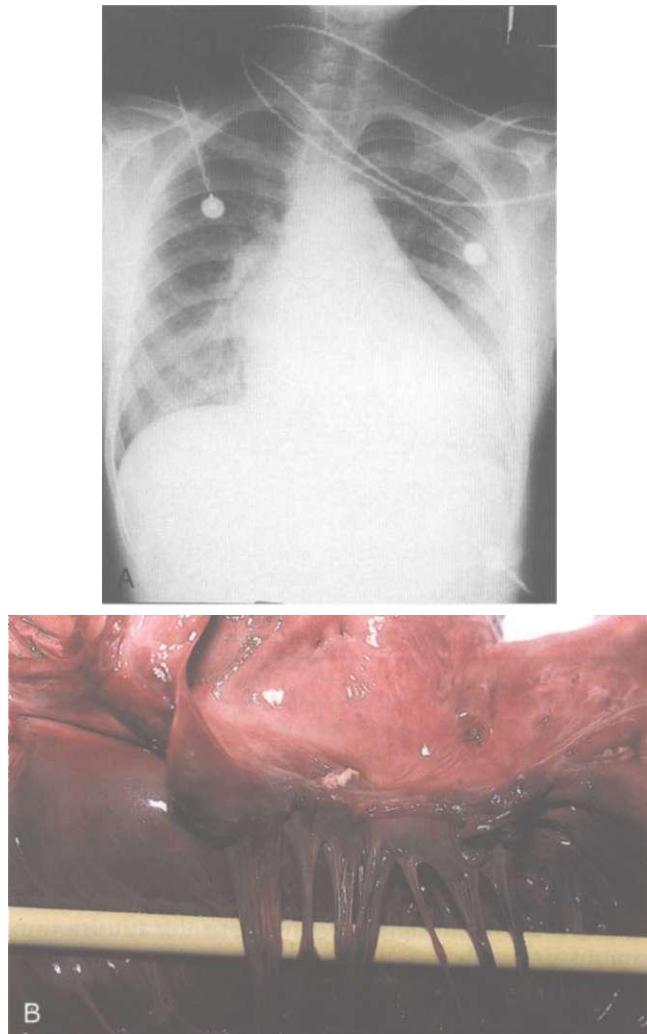


Figure 12-23

A, Chest radiograph of a 15-year-old boy who had multiple occurrences of acute rheumatic fever, showing gross cardiac enlargement and failure. He had mitral regurgitation and stenosis, and aortic regurgitation and stenosis. **B**, Postmortem cardiac examination of the same boy, showing thickened, shortened mitral valve cusps with calcific vegetation and thickened chordae tendineae. Chordae are the tendinous cords connecting the two atrioventricular (AV) valves (the tricuspid valve between the right atrium and right ventricle and the mitral valve between the left atrium and left ventricle) to the appropriate papillary muscles in the heart ventricles; the chordae tendineae in effect anchor the valve leaflets. This support to the AV valves during ventricular systole helps prevent prolapse of the valve into the atrium. (From Cohen J, Powderly WG: *Infectious diseases*, ed 2, St Louis, 2004, Mosby. Courtesy Professor Bart Currie, Darwin, NT, Australia.)

Rheumatic Fever and Heart Disease

Overview, Incidence, and Etiologic Factors

Rheumatic fever is one form of endocarditis (infection), caused by streptococcal group A bacteria, that can be fatal or may lead to rheumatic heart disease (10% of cases), a chronic condition caused by scarring and deformity of the heart valves (Figs. 12-22 and 12-23). It is called rheumatic fever because two of the most common symptoms are fever and joint pain.

The infection generally starts with strep throat in children between ages 5 and 15 years and damages the heart in approximately 50% of cases. The aggressive use of specific antibiotics in the United States had effectively reduced the incidence of rheumatic fever to around 0.5 cases per 100,000 school-age children and removed it as the primary cause of valvular damage.

However, between 1985 and 1987, a series of epidemics of rheumatic fever were reported in several widely

diverse geographic regions of the continental United States, affecting children, young adults aged 18 to 30 years, and, occasionally, middle-aged persons. Currently, the prevalence and incidence of cases have not approximated the 1985 record, but they have remained above previous levels.

Pathogenesis

The exact pathogenesis is unclear, but rheumatic fever produces a diffuse, proliferative, and exudative inflammatory process in the connective tissue of certain struc-

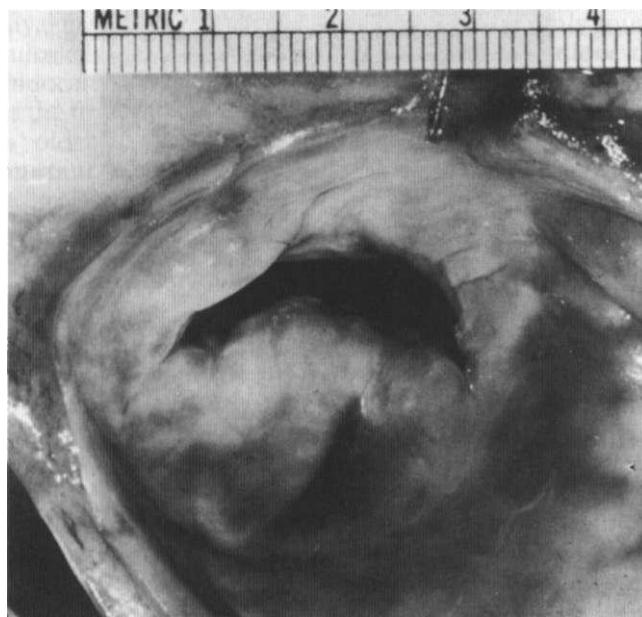


Figure 12-24

When viewed from the atrial aspect, a severely stenotic mitral valve has a narrowed orifice that has the appearance of a classic fish-mouth deformity. (From Kissane JM, ed: *Anderson's pathology*, St Louis, 1990, Mosby.)

tures. The bacteria adhere to the oral and pharyngeal cells and then release their degradation products. Antigens to streptococcal cells bind to receptors on the heart, brain cells, muscles, and joints, which begins the autoimmune response; thus rheumatic fever is classified as an autoimmune disease. In the case of the heart valves, the inflammatory products cross-react with cardiac proteins, affecting cardiac valve tissue and myocardium.

All layers of the heart (epicardium, endocardium, myocardium, pericardium) (see Fig. 12-1) may be involved, including the valves. Endocardial inflammation causes swelling of the valve leaflets, with secondary erosion along the lines of leaflet contact. Small, beadlike clumps of vegetation containing platelets and fibrin are deposited on eroded valvular tissue and on the chordae tendineae; the mitral and aortic valves are most commonly affected.

Chordae are the tendinous cords connecting the two atrioventricular valves (the tricuspid valve between the right atrium and right ventricle and the mitral valve between the left atrium and left ventricle) to the appropriate papillary muscles in the heart ventricles; the chordae tendineae in effect anchor the valve leaflets. This support to the atrioventricular valves during ventricular systole helps prevent prolapse of the valve into the atrium.

Over time, scarring and shortening of the involved structures occur, and the leaflets adhere to each other as the valves lose their elasticity. As many as 25% of clients will have mitral valvular disease 25 to 30 years later, with fibrosis and calcification of valves, fusion of commissures (union or junction between adjacent cusps of the heart valves) and chordae tendineae, and mitral stenosis with fish-mouth deformity (Fig. 12-24).

Clinical Manifestations

Although strep throat is the most common manifestation of the streptococcal virus, streptococcal infections can also affect the skin and, less commonly, the lungs. In some cases of strep throat the initial triggering sore throat or pharyngitis does not cause extreme illness, if any discomfort at all.

However, the major manifestations of acute rheumatic fever are usually carditis, acute migratory polyarthritis, and chorea, which may occur singly or in combination. In the acute, full-blown sequelae, shortness of breath and increasing nocturnal cough also occur. A ring- or crescent-shaped rash with clear centers on the skin of the limbs or trunk (erythema marginatum) is present in fewer than 2% of persons in an acute episode. Subcutaneous nodules may occur over bony prominences and along the extensor surfaces of the arms, heels, knees, or back of the head, but these do not interfere with joint function.

Carditis is most likely to occur in children and adolescents. Mitral or aortic semilunar valve dysfunction (see Pathogenesis) may result in a previously undetected murmur. Chest pain caused by pericardial inflammation and characteristic heart sounds may occur. *Polyarthritis* may develop in a child or young adult with acute rheumatic fever 2 to 3 weeks after an initial cold or sore throat. Sudden or gradual onset of painful migratory joint symptoms in knees, shoulders, feet, ankles, elbows, fingers, or neck; fever (99° to 103° F [37.2° to 39.4° C]); palpitations; and fatigue are present. Malaise, weakness, weight loss, and anorexia may accompany the fever.

The migratory arthralgias usually involve two or more joints simultaneously or in succession and may last only 24 hours, or they may persist for several weeks. In adults, only a single joint may be affected. Joints that are sore and hot and contain fluid completely resolve, followed by acute synovitis, heat, synovial space tenderness, swelling, and effusion present in a different area the next day. The persistence of swelling, heat, and synovitis in a single joint or joints for more than 2 to 3 weeks is extremely unusual in acute rheumatic fever.

Rheumatic *chorea* (also called Sydenham's chorea or St. Vitus' dance) occurs in 3% of cases 1 to 3 months after the streptococcal infection and is always preceded by polyarthritis. Chorea in a child, teenager, or young adult is almost always a manifestation of acute rheumatic fever. Other causes of chorea are SLE, thyrotoxicosis, and cerebrovascular accident, but these are uncommon and unlikely in a child.

The chorea develops as rapid, purposeless, nonrepetitive movements that may involve all muscles except the eyes. This pattern of movement may last for 1 week, several months, or even several years without permanent impairment of the CNS.

MEDICAL MANAGEMENT

DIAGNOSIS AND TREATMENT. Late diagnosis can have serious consequences requiring immediate antibiotic and antiinflammatory treatment. Jones criteria are used as the basis for diagnosis (Table 12-17), and results of throat culture for group A streptococci are usually positive. Echocardiography combined with Doppler technology

Table 12-17 Jones Criteria for Diagnosis of Rheumatic Fever

Major Manifestations	Minor Manifestations	Supporting Evidence of Streptococcal Infection
Carditis	Previous rheumatic fever or rheumatic heart disease	Recent scarlet fever
Polyarthritis	Arthralgia	Positive throat culture results for group A streptococci
Chorea	Fever	Other positive laboratory test results
Erythema marginatum	Elevated level of C-reactive protein	
Subcutaneous nodules	Leukocytosis	
	Electrocardiographic changes	

From Dajani AS, Ayoub A, Burman FZ, et al: American Heart Association medical/scientific statement: guidelines for the diagnosis of rheumatic fever: Jones criteria, 1992 update, *Circulation* 87:302-307, 1993. The Jones criteria have been reviewed and remain valid per the Jones Criteria Working Group. Source: Ferrieri P: Proceedings of the Jones Criteria Workshop, *Circulation* 106:2521-2523, 2002.

provides reliable hemodynamic and anatomic data in the assessment of rheumatic heart disease.

Aspirin may be used to treat the joint manifestations and as a general antiinflammatory agent. Corticosteroids are used when there is clear evidence of rheumatic carditis. Children with acute chorea are generally treated with some form of CNS depressant, such as phenobarbital. Commissurotomy and prosthetic valve replacement may be necessary for valvular dysfunction associated with chronic rheumatic disease.

PROGNOSIS. Initial episodes of rheumatic fever last weeks to months, but 20% of children affected have recurrences within 5 years; relapses increase the risk of heart damage that leads to rheumatic heart disease, with mitral or aortic stenosis or insufficiency caused by progressive valve scarring. Mortality for acute rheumatic fever is low (1% to 2%), but persistent rheumatic activity with complications (enlarged heart, AF, arterial embolism, heart failure, pericarditis) is associated with long-term morbidity and mortality.

enough that the person does not seek medical care of any kind. The presence of fever accompanied by a clinical presentation of migratory arthralgias or a history of recent illness as described requires medical evaluation.

The risk of developing acute rheumatic fever following untreated tonsillopharyngitis is only 1% in the pediatric population, but as many as 25% of people affected by rheumatic fever develop mitral valve dysfunction 25 to 30 years later. Adults who experience exercise intolerance or exertional dyspnea of unknown cause and who have a previous history of childhood rheumatic fever may be experiencing the effects of MVP. Dyspnea associated with MVP is most commonly accompanied by fatigue and palpitations. This history in combination with this triad of symptoms requires evaluation by a physician.

In the case of a confirmed diagnosis of rheumatic fever-related mitral valve involvement, exercise will not improve the mechanical function of the valve, but improvement in cardiovascular function can occur. (See Special Implications for the Therapist: Valvular Heart Disease.)

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-15

Rheumatic Fever and 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

Other practice patterns may apply in the presence of polyarthritis or chorea.

The increased incidence of infection with streptococcal group A bacteria in the adult population may result in cases of sudden or gradual onset of painful migratory joint symptoms affecting the knees, shoulders, feet, ankles, elbows, fingers, or neck. Any time an adult presents with intermittent or migratory joint symptoms, the client's temperature must be taken.

The therapist should ask about recent exposure to someone with strep throat and a recent history or presence of rash anywhere on the body, sore throat, or cold. The sore throat or cold symptoms may be mild

DISEASES AFFECTING THE PERICARDIUM

The pericardium consists of two layers: the inner visceral layer, which is attached to the epicardium; and an outer parietal layer (see Fig. 12-1). The pericardium stabilizes the heart in its anatomic position despite changes in body position and reduces excess friction between the heart and surrounding structures. It is composed of fibrous tissue that is loose enough to permit moderate changes in cardiac size but that cannot stretch fast enough to accommodate rapid dilation or accumulation of fluid without increasing intracardiac pressure.

The pericardium may be a primary site of disease and is often involved by processes that affect the heart; it may also be affected by diseases of the adjacent tissues. Pericardial diseases are common and have multiple causes. Three conditions primarily affect the pericardium: acute pericarditis, constrictive pericarditis, and pericardial effu-

Box 12-12**CAUSES OF PERICARDITIS**

- Idiopathic (85%)
- Infections
 - Viral (Coxsackie, influenza, Epstein-Barr, hepatitis, human immunodeficiency virus)
 - Bacterial (tuberculosis, *Staphylococcus*, *Streptococcus*, *Meningococcus*, pneumonia)
 - Parasitic
 - Fungal
- Myocardial injury
 - Myocardial infarction (MI)
 - Cardiac trauma: instrumentation; blunt or penetrating pericardium; rib fracture
 - Post cardiac surgery
- Hypersensitivity
 - Collagen diseases: rheumatic fever, scleroderma, systemic lupus erythematosus (SLE), rheumatoid arthritis
 - Drug reaction
 - Radiation or cobalt therapy
- Metabolic disorders
 - Uremia
 - Myxedema
- Chronic anemia
- Neoplasm
 - Lymphoma, leukemia, lung or breast cancer
- Aortic dissection
- Graft-versus-host disease

Pathogenesis

Many causes of pericarditis affect both the pericardium and the myocardium (myopericarditis) with varying degrees of cardiac dysfunction. Constrictive pericarditis is characterized by a fibrotic, thickened, and adherent pericardium that is compressing the heart. The heart becomes restricted in movement and function (cardiac tamponade). Diastolic filling of the heart is reduced, venous pressures are elevated, cardiac output is decreased, and eventual cardiac failure may result.

When fluid accumulates within the pericardial sac it is referred to as *pericardial effusion*. Blunt chest trauma or any cause of acute pericarditis can lead to pericardial effusion. Rapid distention or excessive fluid accumulation from this condition can also compress the heart and reduce ventricular filling and cardiac output.

Clinical Manifestations

The presentation and course of pericarditis are determined by the underlying etiology. For example, pericarditis may occur 2 to 5 days after infarction as a result of an inflammatory reaction to myocardial necrosis, or it may occur within the first year after radiation initiates a fibrinous and fibrotic process in the pericardium. Often there is pleuritic chest pain that is made worse by lying down and by respiratory movements and is relieved by sitting upright or leaning forward. The pain is substernal and may radiate to the neck, shoulders, upper back, upper trapezius, left supraclavicular area, or epigastrium, or down the left arm.

Other symptoms may include fever, joint pain, dyspnea, or difficulty swallowing. Auscultation of the lower left sternal border where the pericardium lies close to the chest wall will produce a pericardial friction rub, a high-pitched scratchy sound that may be heard at end expiration. This sound is produced by the friction between the pericardial surfaces that results from inflammation and occurs during heart movement. Symptoms of constrictive pericarditis develop slowly and usually include progressive dyspnea, fatigue, weakness, peripheral edema, and ascites. Constrictive disease can lead to diastolic dysfunction and eventual heart failure.

MEDICAL MANAGEMENT

DIAGNOSIS. Clinical examination, including clinical presentation, auscultation, and client history, may be diagnostic. A classic sign of pericarditis is the pericardial friction rub heard on auscultation. Other diagnostic tools include chest x-ray (showing enlarged cardiac shadow), characteristic ECG changes (showing evidence of an underlying inflammatory process), and laboratory studies (e.g., elevated sedimentation rate or elevated white blood count [nonspecific indicators of inflammation] and elevated cardiac enzymes [post MI]). CT, MRI, and echocardiography are modalities used for imaging the pericardium and pericardial disease.

TREATMENT. New treatments for pericardial diseases are being developed as a result of modern imaging, new understanding of molecular biology, and immunologic techniques. Comprehensive and systematic implementa-

sion. These three diseases are grouped together for ease of understanding in the following section.

Pericarditis**Definition and Overview**

Pericarditis or inflammation of the pericardium, the double-layer membrane surrounding the heart, may be a primary condition or may be secondary to a number of diseases and circumstances (Box 12-12). It may occur as a single acute event, or it may recur and become a chronic condition called *constrictive pericarditis* (uncommon).

Incidence and Etiologic Factors

The most common types of pericarditis encountered by the therapist will be drug induced or those present in association with autoimmune diseases (e.g., connective tissue disorders such as SLE, rheumatoid arthritis), after MI, in conjunction with renal failure, after open heart surgery, and after radiation therapy.

Other types encountered less often include viral pericarditis (e.g., Epstein-Barr, hepatitis, human immunodeficiency virus [HIV]) and neoplastic pericarditis (from spread to the pericardium of adjacent lung cancer or invasion by breast cancer, leukemia, Hodgkin's disease, or lymphoma). Isolated cases of constrictive pericarditis as a manifestation of chronic graft-versus-host disease after peripheral stem cell transplantation have been reported.³⁰⁹

tion of new techniques of pericardiocentesis, pericardial fluid analysis, pericardioscopy, and epicardial and pericardial biopsy, as well as the application of new techniques for pericardial fluid and biopsy analyses, have permitted early specific diagnosis, creating foundations for etiologic intervention in many cases. In cases of recurrent pericarditis resistant to conventional intervention and in the case of neoplastic pericarditis, intrapericardial application of medication has been proposed.²⁰⁷

Conventional treatment remains twofold, directed toward prevention of long-term complications and the underlying cause. For example, while any underlying infection is treated when possible (antibiotics for bacterial pericarditis), symptomatic treatment is provided for idiopathic, viral, or radiation pericarditis; antiinflammatory drugs are given for severe, acute pericarditis or pericarditis associated with connective tissue disorders; chemotherapy is given for neoplastic pericarditis; and dialysis is performed for uremic pericarditis. Analgesics may be prescribed for the pain and fever. Pericardiocentesis (surgical drainage with a needle catheter through a small subxiphoid incision) may be performed if cardiac compression from pericardial effusion does not resolve.

Treatment for constrictive pericarditis is both medical and surgical, including digitalis preparations, diuretics, sodium restriction, and pericardectomy (surgical excision of the damaged pericardium).

PROGNOSIS. The prognosis in most cases of acute viral pericarditis is excellent when there is no (or only minimal) myocardial involvement, since this is frequently a self-limited disease. Without medical intervention, shock and death can occur from decreased cardiac output with cardiac involvement. Constrictive pericarditis is a progressive disease without spontaneous reversal of symptoms. Most people become progressively disabled over time. Surgical removal of the pericardium is associated with a high mortality rate when progressive calcification in the epicardium and dense adhesions or fibrosis between the pericardial layers are present.

Special precautions depend on the underlying cause of the pericarditis. Mild cases require intervention per client tolerance, and the therapist observes for any symptoms of CHF. A mild pericarditis can quickly progress to a severe condition that requires medical evaluation. The clinician is referred to each individual section in this text representing the etiology of pericarditis for precautions.

DISEASES AFFECTING THE BLOOD VESSELS

Diseases of blood vessels observed in a therapy setting can include intestinal infarction, aneurysm, PVD, vascular neoplasm, and vascular malformation; only intestinal infarction will not be discussed here.

Aneurysm

Definition and Overview

An aneurysm is an abnormal stretching (dilation) in the wall of an artery, a vein, or the heart with a diameter that is at least 50% greater than normal. When the vessel wall becomes weakened from trauma, congenital vascular disease, infection, or atherosclerosis, a permanent saclike formation develops. A false aneurysm can occur when the wall of the blood vessel is ruptured and blood escapes into surrounding tissues, forming a clot (Fig. 12-25; see also Fig. 12-27).

Aneurysms are of various types (either arterial or venous) and are named according to the specific site of formation (Fig. 12-26). The most common site for an arterial aneurysm is the aorta, forming a thoracic aneurysm (which involves the ascending, transverse, or first part of the descending portion of the aorta) or an abdominal aneurysm (which generally involves the aorta between the renal arteries and iliac branches).

Thoracic aortic aneurysms located above the diaphragm account for approximately 10% of all aortic aneurysms and occur most frequently in hypertensive men between the ages of 40 and 70 years. Men are more likely to have thoracic or abdominal aneurysms. Thoracic aortic aneurysms occur less often than other types but tend to be more life-threatening.

Abdominal aortic aneurysms located below the diaphragmatic border occur about four times more often than thoracic aneurysms, most likely because the aorta is not supported by skeletal muscle at this location. The incidence of abdominal aortic aneurysm is increasing, probably because of the increasing number of adults over 65 years of age.

Peripheral arterial aneurysms affect the femoral and popliteal arteries.

Incidence and Etiologic Factors

According to the Society for Vascular Surgery, approximately 200,000 people in the United States are diagnosed annually with aortic aneurysm and 15,000 of those aneu-

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-16

Pericarditis

PREFERRED PRACTICE PATTERN

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

Pericardial pain can masquerade as a musculoskeletal problem, presenting as just upper back, neck, or upper trapezius pain. In such cases, the pain may be diminished by holding the breath or aggravated by swallowing or neck or trunk movements, especially side bending or rotation.

Pain is also aggravated by respiratory movements, such as deep breathing, coughing, and laughing. The therapist must screen for medical disease by assessing aggravating and relieving factors and by asking the client about a history of fever, chills, upper respiratory tract infection (recent cold or flu), weakness, heart disease, or recent MI (heart attack).

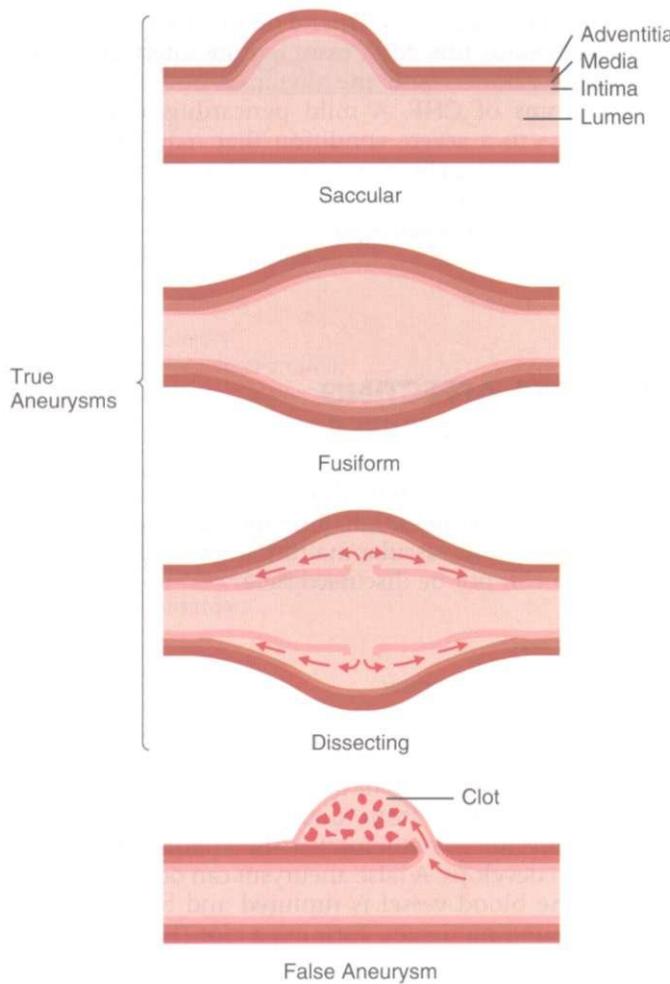


Figure 12-25

Longitudinal sections showing types of aneurysms. In a true aneurysm, layers of the vessel wall dilate in one of the following ways: saccular, a unilateral outpouching; fusiform, a diffuse dilation involving the entire circumference of the artery wall; or dissecting, a bilateral outpouching in which layers of the vessel wall separate, with creation of a cavity. In a false aneurysm, the wall ruptures, and a blood clot is retained in an outpouching of tissue.

rysms are severe enough to rupture, causing a medical emergency.³¹²

Incidence increases with increasing age, usually beginning after age 50 years, presumably as a result of chronic inflammatory cellular changes resulting in atherosclerosis. However, someone without evidence of atherosclerosis can develop an aneurysm, especially in the presence of congenital weakness of the blood vessel walls.

Family members (parent, adult child, or sibling) of anyone with an aneurysm have a fourfold increased risk of aneurysm, and gene defects on chromosomes 11³⁴⁰ and 15¹⁷² have been identified with some of the connective tissue disorders associated with aneurysm. Recently, a mutation in a specific protein, transforming growth factor β receptor (TGFBR), has been identified as responsible for causing aneurysms.²⁰⁰ Aneurysms occur much more often in men than in women, and one half of affected persons are hypertensive.

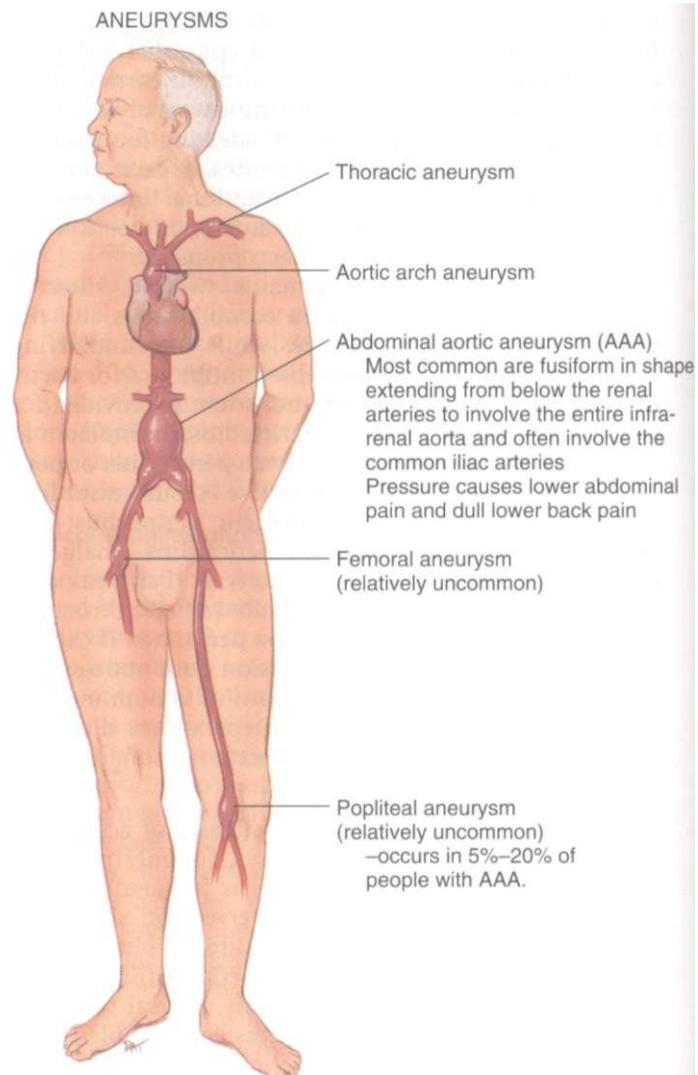


Figure 12-26

Aneurysms are named according to the specific site of formation. Abdominal aortic aneurysms are the most common type; more than 95% of abdominal aortic aneurysms are located below the renal arteries and extend to the umbilicus, causing low back pain. (From Jarvis C: *Physical examination and health assessment*, ed 5, Philadelphia, 2008, Saunders.)

Atherosclerosis or any injury to the middle or muscular layer of the arterial wall (tunica media) is responsible for most arterial aneurysms. Other less common causes of aneurysm include trauma (blunt or surgical), Marfan's disease (congenital defects of the arterial wall) and other hereditary abnormalities of connective tissue, and inflammatory diseases and infectious agents (bacterial infection, syphilis, polyarteritis).

The emergence of HIV has been associated with a dramatic increase in the incidence of syphilis. Since syphilitic aortitis generally presents between 10 and 30 years after the primary infection, there may be an increased incidence of associated aneurysms in the coming years. Hypertension seems to enhance aneurysm formation.

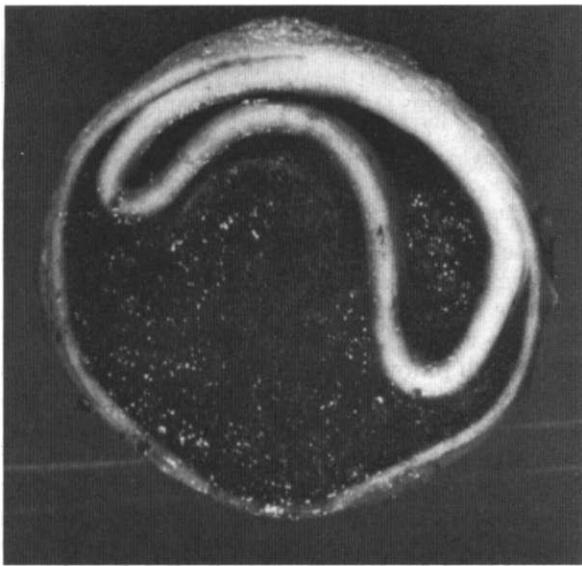


Figure 12-27

Dissecting aneurysm. Cross-section of the aorta with dissecting aneurysm showing true aortic lumen (above and right) compressed by dissecting column of blood that separates the media and creates a false lumen. [From Kissane JM, ed: *Anderson's pathology*, St Louis, 1990, Mosby.]

Pathogenesis

Plaque formation erodes the vessel wall, predisposing the vessel to stretching of the inner and outer layers of the artery and formation of a sac. The stretching of the media produces infarct expansion, a weak and thin layer of necrotic muscle, and fibrous tissue that bulges with each systole. Abnormal proteolysis, the presence of elastolytic serum enzymes, and deficiencies of collagen and elastin have been implicated as factors contributing to the development of these aneurysms.¹⁸⁴

With time, the aneurysm becomes more fibrotic, but it continues to bulge with each systole, thus acting as a reservoir for some of the stroke volume. In the case of thoracic aortic aneurysms, the shear force of elevated blood pressure causes a tear in the intima with rapid disruption and rupture of the aortic wall. Subsequent hemorrhage causes a lengthwise splitting of the arterial wall, creating a false vessel (Fig. 12-27), and a hematoma may form in either channel (i.e., the false or true lumen).

Clinical Manifestations

Aneurysms may be asymptomatic; when they do occur, manifestations depend largely on the size and position of the aneurysm and its rate of growth. Persistent but vague substernal, back, neck, or jaw pain may occur as enlargement of the aneurysm impinges adjacent structures.

Dissection over the aortic arch and into the descending aorta may be experienced as extreme, sharp pain felt at the base of the neck or along the back into the interscapular areas. When pressure from a large volume of blood is placed on the trachea, esophagus, laryngeal nerve, lung,

or superior vena cava, symptoms of dysphagia; hoarseness; edema of the neck, arms, or jaw and distended neck veins; and dyspnea and/or cough may occur, respectively.

Other signs and symptoms may be present in the case of *acute aortic dissection* as a result of compression of branches of the aorta. These include acute MI, reversible ischemic neurologic deficits, stroke, paraplegia, renal failure, intestinal ischemia, and ischemia of the arms and legs. Acute chest pain may also result from a nondissecting hematoma of the aorta or erosion of a penetrating atherosclerotic ulcer.¹⁸⁴

In the case of an untreated *abdominal aortic aneurysm* expansion and rupture can occur in one of several places, including the peritoneal cavity, the mesentery, the retroperitoneum, into the inferior vena cava, or into the duodenum or rectum. Rupture refers to a tearing of all three tunicae (tunica adventitia, tunica media, tunica intima) with bleeding into the thoracic or abdominal cavity. The most common site for an abdominal aortic aneurysm is just below the renal arteries, and it may involve the bifurcation of the aorta (see Fig. 12-26).

Most abdominal aortic aneurysms are asymptomatic, but intermittent or constant pain in the form of mild to severe mid-abdominal or lower back discomfort is present in some form in 25% to 30% of cases. Groin or flank pain may be experienced because of increasing pressure on other structures.

Early warning signs of an impending rupture may include abdominal heartbeat when lying down or a dull ache (intermittent or constant) in the mid-abdominal left flank or lower back. Rupture is most likely to occur in aneurysms that are 5 cm or larger, causing intense flank pain with referred pain to the back at the level of the rupture. Pain may radiate to the lower abdomen, groin, or genitalia. Back pain may be the only presenting symptom before rupture occurs.

The most common site for *peripheral arterial aneurysm* is the popliteal space in the lower extremities. Most are caused by atherosclerosis and occur bilaterally in men. Popliteal aneurysm presents as a pulsating mass, 2 cm or more in diameter, and causes ischemic symptoms in the lower limbs (e.g., intermittent claudication, rest pain, thrombosis and embolization resulting in gangrene). *Femoral aneurysm* presents as a pulsating mass in the femoral area on one or both sides.

MEDICAL MANAGEMENT

DIAGNOSIS. Detection of abdominal and peripheral aneurysms often occurs when the physician palpates a pulsating mass during routine examination or when x-rays are taken for other purposes (although not all aortic aneurysms show abnormalities on chest radiography). Radiography, ultrasonography, echocardiography with color Doppler imaging, CT and MRI, arteriography, and aortography may be used for investigation.

PREVENTION AND TREATMENT. Annual examination to ensure early identification is recommended for family members (parent, adult child, or sibling) of anyone who has previously been diagnosed with an aortic aneurysm. Anyone with a family risk or signs of diseased arteries

should take preventive measures, including smoking cessation, regular exercise, blood pressure control, and cholesterol management.

Treatment is determined based on the size of the bulge, how fast it is expanding, and the individual's clinical presentation. For small aneurysms, watchful waiting is often advised. Preventive pharmacology (e.g., statin to lower cholesterol, β -blocker or ACE inhibitor to control blood pressure) may be prescribed depending on individual factors.

Surgical intervention before rupture provides a good prognosis; at 5 cm, the risk of rupture exceeds the risk of repair. A new, less invasive procedure known as *endoluminal stent-graft* may offer an alternative to open abdominal surgery. Guided by angiographic imaging, a catheter is inserted through the femoral or brachial artery to the aneurysm. A balloon within the catheter is then inflated, pushing open the stent, which attaches with tiny hooks to healthy arterial wall above and below the aneurysm. This creates a channel for blood flow that bypasses the aneurysm.

PROGNOSIS. The standard open surgical approach to replace the diseased aorta is steadily improving but is still associated with high morbidity and substantial mortality rates. MI, respiratory failure, renal failure, and stroke are the principal causes of death and morbidity after surgical procedures performed on the thoracic aorta.

At the same time, the endoluminal stent-graft comes with its own set of complications, including fever, breakdown or migration of the device, leaks, and unknown durability. Further studies to improve treatment are ongoing. Aneurysm rupture is associated with a high mortality; frequently, aneurysms are discovered only at autopsy.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-17

Aneurysm

PREFERRED PRACTICE PATTERNS

4J: Impaired Motor Function, Muscle Performance, Range of Motion, Gait, Locomotion, and Balance Associated with Amputation (peripheral aneurysm)

5D: Impaired Motor Function and Sensory Integrity Associated with Nonprogressive Disorders of the Central Nervous System—Acquired in Adolescence or Adulthood

5I: Impaired Arousal, Range of Motion, and Motor Control Associated with Coma, Near Coma, or Vegetative State

Anyone with complications associated with an aneurysm is also at risk for pulmonary complications.

Since the prevalence of all diseases of the aorta increases with age and because the population in the United States is aging, it is expected that aortic aneurysm will be encountered with increasing frequency. Knowledge of the natural history, familial history, and clinical features of this disorder may alert the therapist to the need for medical intervention.

For the person who has had a surgically repaired aneurysm, activities are restricted and are only gradu-

ally reintroduced. The therapist may be involved in bedside exercises and early mobility, which are especially important to prevent thromboembolism as a result of venous stasis during prolonged bed rest and immobility.

Because of the invasiveness of open abdominal surgery, anyone undergoing this procedure is at high risk for pulmonary complications. Incisional pain and the use of abdominal musculature in coughing discourage the person from full inspirations as well as effective forceful huffing or coughing. The acute care therapist will utilize clinical techniques to assist with cough with pillows or towel rolls at the incisional site and forceful huffing (see description in Chapter 15¹⁵⁰).

Proper lifting techniques should be reviewed before discharge, even though the client will not be able to provide a return demonstration. Activities that require pushing, pulling, straining, or lifting more than 10 lb are restricted for 6 to 10 weeks postoperatively.

Anterior or abdominal soft tissue mobilization for persons with back pain who have postoperative abdominal scars may require indirect techniques. This precaution is especially true for the person with a previous abdominal aneurysm, the person with a known nonoperative aneurysm (less than 5 cm), or the person with a family history of aneurysm or an undiagnosed aneurysm.

The therapist must always palpate the abdomen for a pulsating mass before performing anterior or abdominal therapy. It is possible to palpate the width of the pulse beginning at the abdominal midline and progressing laterally. The pulse should be characterized by a uniform width on either side of the abdominal midline until the umbilicus is reached, at which point the aortic bifurcation results in expansion of the pulse width. Throbbing pain that increases with exertion should alert the therapist to the need to monitor vital signs and palpate pulses.

Peripheral Vascular Disease

Although PVD is usually thought to refer to diseases of the blood vessels supplying the extremities, in fact, PVD actually encompasses pathologic conditions of blood vessels supplying the extremities and the major abdominal organs, most often apparent in the intestines and kidneys.

PVD is organized based on the underlying pathologic finding (e.g., inflammatory, arterial occlusive, venous, or vasomotor disorders) (Box 12-13). Although the terms *peripheral arterial disease* (PAD) and *peripheral vascular disease* (PWD) are often used interchangeably, PVD is a broader, more encompassing grouping of disorders of both the arterial and venous blood vessels, whereas PAD only refers to arterial blood vessels. PVD typically affects the legs more often than the arms, but upper extremity involvement is not uncommon.

Approximately 8 million Americans over age 60 years are affected by PVD, with 20% of those people over age

Box 12-13**PERIPHERAL VASCULAR DISEASES**

Inflammatory Disorders	Arterial Occlusive Disorders	Venous Disorders	Vasomotor Disorders
<ul style="list-style-type: none"> • Vasculitis (see also Table 12-18) • Polyarteritis nodosa • Arteritis • Allergic or hypersensitivity angiitis • Kawasaki disease • Thromboangiitis obliterans (Buerger's disease) 	<ul style="list-style-type: none"> • Arterial thrombosis/embolism • Thromboangiitis obliterans • Arteriosclerosis obliterans 	<ul style="list-style-type: none"> • Thrombophlebitis • Varicose veins • Chronic venous insufficiency 	<ul style="list-style-type: none"> • Raynaud's disease • Complex regional pain syndrome (CRPS, formerly reflex sympathetic dystrophy [RSD])

70 years. Like CAD and cerebrovascular disease, arterial occlusive forms of PVD are most common as a result of atherosclerosis. Intermittent claudication is the classic symptom of PAD. Like angina associated with CAD, intermittent claudication associated with PAD is predictable and nearly always develops after the same amount of exertion (e.g., walking a specific distance), generally occurs in the calves and less commonly in the thighs and buttocks,⁶⁸ and usually improves rapidly with rest.

Data from the Framingham Heart Study and other population studies indicate that intermittent claudication sharply increases in late middle age and is somewhat higher among men than women. In fact the true prevalence of PAD is at least five times higher than expected based on the reported prevalence of intermittent claudication.⁶⁷ Specific symptoms of the various forms of PVD depend on the underlying pathologic condition, the blood vessels involved (arteries or veins), and the location of the affected blood vessels; each form is discussed individually in the following sections.

Inflammatory Disorders

Inflammatory conditions of the blood vessels are often discussed as immunologic conditions, because inflammation and damage to large and small vessels result in end-stage organ damage. Vasculitis (e.g., arteritis, such as polyarteritis nodosa and giant cell arteritis, Kawasaki disease) is the most commonly encountered inflammatory blood vessel disease in a therapy practice.

Vasculitis is actually a group of disorders that share a common pathogenesis of inflammation of the blood vessels involving arteries, veins, or nerves, resulting in narrowing or occlusion of the lumen or formation of aneurysms that can rupture. Vascular inflammation is a central feature of many rheumatic diseases, especially rheumatoid arthritis and scleroderma. (See also the section on Rheumatoid Vasculitis in Chapter 5.)

Vasculitis. Vasculitis can involve blood vessels of any size, type, or location and can affect any organ system, including the nervous system; classification is usually according to the size of the predominant vessels involved (Table 12-18). Vasculitis may be acute or chronic with varying degrees of involvement. The distribution of lesions may be irregular and segmental rather than continuous.

Neurologic manifestations of vasculitis can occur in conjunction with any of the vasculitides listed, affecting

the peripheral nervous system or the CNS. Vasculitis may occur as an isolated peripheral nerve vasculitis (localized vasculitis). Numerous vasculitic diseases have been reported in association with HIV disease. The primary target organ involvement is usually muscle and nerve, skin, testicle, kidney, and, less often, the CNS.

Immune (antibody-antigen) complexes to each disorder are deposited in the blood vessels, resulting in varying symptoms depending on the organs affected. In the case of vasculitic neuropathy, the formation of antibody-antigen complexes activates the complement cascade with generation of C3a and C5a (chemotactic agents that recruit polymorphonuclear [PMN] leukocytes to the vessel walls).

Phagocytosis of the immune complexes takes place, and release of free radicals and proteolytic enzymes disrupts cell membranes and damages blood vessel walls. The complement cascade generates the formation of complement membrane attack complex that also contributes to endothelial damage (see discussion in Chapter 6; see also Fig. 6-15). The resulting damage to endothelial cells results in thickening of the vessel wall, occlusion, and ischemia of the affected nerves with axonal degeneration and the resultant neuropathy.

Polyarteritis Nodosa

Overview and Etiologic Factors. Polyarteritis nodosa refers to a condition consisting of multiple sites of inflammatory and destructive lesions in the arterial system; the lesions are small masses of tissue in the form of nodes or projections (nodosum). The cause of polyarteritis nodosa is unknown, although hepatitis B is present in 50% of cases, and polyarteritis occurs more commonly among IV drug abusers and other groups who have a high prevalence of hepatitis B (see the section on Hepatitis B in Chapter 17). Any age can be affected, but it is more common among young men.

Clinical Manifestations. Polyarteritis nodosa affects small and medium-sized blood vessels, resulting in a variety of clinical presentations depending on the specific site of the blood vessel involved. Some of the more likely symptoms include abrupt onset of fever, chills, tachycardia, arthralgia, and myositis with muscle tenderness.

Any organ of the body may be affected, but most often involved are the kidneys, heart, liver, GI tract, muscles, and testes. Abdominal pain, nausea, and vomiting are common with GI tract involvement. Pericarditis, myocarditis, arrhythmias, and MI reflect cardiac involvement.

Table 12-18 Vasculitis

Vessels Involved	Vasculitides	Organ Systems Involved
Small vessels (arterioles, capillaries, venules)	Inflammatory bowel disease vasculitis	Skin, viscera, heart, synovium, GI tract
	Hypersensitivity vasculitis; drug-induced vasculitis	According to underlying cause and involved structures
	Vasculitis associated with infections or other diseases	
	Immune-complex small vessel vasculitis associated with autoimmune conditions (paraneoplastic vasculitis)	
Medium-sized and small muscular vessels	Vasculitis associated with malignancy	Determined by site of malignancy
	Thromboangiitis obliterans (Buerger's disease)	Arteries and veins of digits and limbs
	Kawasaki disease (muscular arteries, rarely veins)	Cardiac, iliac, renal, internal mammary vessels
	Polyarteritis nodosa	Aorta and its primary and secondary branches, renal and visceral arteries, muscles, testes, nerves
Large and medium-sized vessels	Vasculitis in rheumatic disease and connective tissue disorders	Synovium, skin, nail beds
	Angitis of the CNS	CNS
	Wegener's granulomatosis (uncommon)	Local: nasal structures Systemic: lungs (upper and lower respiratory tracts), kidneys (glomerulonephritis), any organ can be involved
Peripheral nervous system	Takayasu's arteritis (rare)	Aorta and its primary branches, renal and visceral arteries
	Giant cell (temporal) arteritis	Extracranial arteries of the head and neck; any other artery but less common
	Localized vasculitis	Vasa nervorum at the level of the epineurial arteries (i.e., blood vessels supplying the neural arch in the spinal axis)

GI, Gastrointestinal; CNS, central nervous system.

Complications may include aneurysm, hemorrhage, thrombosis, and fibrosis leading to occlusion of the lumen. Multiple asymmetric neuropathies (motor and sensory distribution) can occur when vasculitis affects the arteries of the peripheral nerves (vasa nervorum). Paresthesias, pain, weakness, and sensory loss occur, involving several or many peripheral nerves simultaneously.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Diagnosis is made by characteristic laboratory findings, biopsy of symptomatic sites (especially muscle or nerve), and, possibly, visceral angiography. When CNS vasculitis is suspected, angiography is necessary, because MRI and CT do not provide sufficient evidence to confirm the diagnosis.

Prolonged use of corticosteroids is necessary to control fever and constitutional symptoms while vascular lesions are healing. Immunosuppressants may be used in conjunction with steroids to improve survival; withdrawal from drugs is often followed by relapse. Treatment of polyarteritis nodosa associated with hepatitis B is more complicated, because cytotoxic drugs used to treat the vasculitis can exacerbate the hepatic disease. Prognosis is poor without intervention, with a 5-year survival rate of only 20%. Pharmacologic therapy with corticosteroids increases survival to 50%, and steroids combined with

immunosuppressive drugs have improved 5-year survival to 90%.

Arteritis

Overview and Incidence. Arteritis, sometimes called giant cell arteritis (CCA) or cranial or temporal arteritis, is a vasculitis primarily involving multiple sites of temporal and cranial arteries (i.e., arteries of the head and neck and sometimes the aortic arch).

It is the most common vasculitis in the United States and affects older people; the incidence increases with age after 50 years. Postmenopausal women are affected twice as often as men, especially those individuals who have an arthritis-related disease called *polymyalgia rheumatica* (PMR). Other risk factors identified in women include heart murmurs and smoking.

Etiologic Factors and Pathogenesis. Most studies have shown an association of CCA with a specific human leukocyte antigen (HLA) allele, and tumor necrosis factor appears to influence the susceptibility to both CCA and PMR. There may be an infectious origin, but additional studies are necessary to clarify the genetic influence on susceptibility to these conditions. The molecular pathogenesis of GCA involves interleukin-1, intercellular adhesion molecules, and other factors, but the exact pathogenesis remains unknown. The possible role of female sex hormones requires further investigation.

Immunologic research indicates an antigen-driven disease with local T-cell and macrophage activation in vessel walls with calcified atrophic media. The middle layer (tunica media) of the large and medium-sized arteries, particularly those blood vessels supplying blood to the head, is inflamed, causing the arteries to swell and obstruct blood flow (stenosis); ischemic complications and secondary thrombosis may occur. Healing produces fibrosis of the arterial wall and the affected blood vessel becomes cordlike, thickened, and nodular, which can be observed externally when the temporal artery is involved.

Clinical Manifestations. The onset of arteritis is usually sudden, with severe, continuous, unilateral, throbbing headache and temporal pain as the first symptoms, with flulike symptoms or visual disturbances. The pain may radiate to the occipital area, face, or side of the neck. Visual disturbances range from blurring to diplopia to visual loss. Irreversible blindness may occur anywhere in the course of the disease from involvement of the ophthalmic artery.

Other symptoms may include enlarged, tender temporal artery; scalp sensitivity; and jaw claudication (i.e., pain in response to chewing, talking, or swallowing) when involvement of the external carotid artery causes ischemia of the masseter muscles; the pain is relieved by rest.

Approximately 40% of cases present with nonclassic symptoms of respiratory tract problems (most often dry cough), fever of unknown origin, painful paralysis of a shoulder (mononeuritis multiplex), or claudication of the arm with cold hands, arm weakness, and absent radial pulses.¹⁷⁷ Left untreated, the condition may lead to blindness and occasionally to a stroke, heart attack, or aortic dissection.

MEDICAL MANAGEMENT

DIAGNOSIS. Early diagnosis is important to prevent blindness caused by obstruction of the ophthalmic arteries. Diagnosis is made by recognition of the presenting symptoms, and in some cases, arteritis follows PMR, a similar condition. Although GCA and PMR may occur as separate entities, most epidemiologic surveys group these two conditions together as one disorder. These may be two forms of a common pathophysiologic process characterized by varying degrees of synovitis and arteritis. They may actually represent two points along a single disease continuum.

Biopsy of the temporal artery may be performed, but results are often negative given the focal (segmental) nature of the disease. Color ultrasonography of the temporal arteries detects characteristic signs of vasculitis with a high sensitivity and specificity even in the absence of clinical signs of vascular inflammation (helpful in diagnosing temporal arteritis in people with previously diagnosed PMR).

People with extracranial GCA present with occlusive arterial lesions that may be detected with multiple imaging modalities: arteriography, IV digital subtraction angiography (IV-DSA), CT scanning, and MRA. However, inflammation of the arterial wall cannot be detected by these means.

Standard CT imaging with contrast enhancement and certain MR sequences as well as ultrasound permit identification of the edema and inflammation of the vessel wall. This is an important marker for active disease. Laboratory findings include elevated erythrocyte sedimentation rate, reflective of the underlying inflammatory process.

TREATMENT AND PROGNOSIS. Treatment of arteritis to prevent blindness and other vascular complications is with oral antiinflammatory drugs (usually a corticosteroid such as prednisone), providing symptomatic relief in 3 to 5 days. Visual loss can be permanent if allowed to persist for several hours without adequate intervention. With proper intervention, arteritis is a self-limiting disease, usually resolving within 6 to 12 months. About 30% of affected individuals relapse in the first year of treatment during dose tapering. Alternative therapies of combined pharmacology with corticosteroids and methotrexate may be more effective in controlling disease with fewer complications.¹⁷⁸

Hypersensitivity Angitis. Hypersensitivity angiitis, a form of vasculitis, can occur at any age, but it most commonly affects children and young adults. The etiology is unknown, but the disease often follows an upper respiratory tract infection, and allergy or drug sensitivity plays a role in some cases. It is usually localized to the small vessels of the skin, first appearing on the lower extremities in a variety of possible lesions.

A classic triad of symptoms occurs in 80% of cases that includes purpura (bruising and petechiae or round purplish red spots under the skin), arthritis, and abdominal pain. Inflammation and hemorrhage may occur in the synovium and CNS. Medical management (diagnosis, treatment, prognosis) is the same as for the other forms of vasculitis already discussed.

Kawasaki Disease

Overview and Etiologic Factors. Kawasaki disease, also known as mucocutaneous lymph node syndrome, is an acute febrile illness associated with systemic (multiorgan) vasculitis. It can occur in any ethnic group but seems most prevalent in Asian populations (especially Japanese, with equal incidence in Japan and in the United States among Japanese or Japanese descendants).

Children under the age of 5 years comprise 80% of all cases, and 20% develop cardiac complications that can be fatal. The etiology is unknown, but because seasonal and geographic outbreaks appear to occur, an infectious cause is suspected. Current etiologic theories center on an immunologic response to an infectious, toxic, or antigenic substance.¹⁹⁷

Pathogenesis. Substantial evidence suggests that immune activation has a role in the pathogenesis of Kawasaki syndrome. The principal area of pathologic findings is the cardiovascular system. Kawasaki disease progresses pathologically and clinically in stages. During the acute stage of the illness (first 2 weeks) vascular inflammation and immune activation within the arterioles, venules, and capillaries occur, which later progress (stage 2, weeks 2 to 4) to include the main coronary arteries, the heart, and the larger veins.

The acute phase is also associated with the appearance of circulating antibodies that are cytotoxic against vascular endothelial cells; the presence of elevated antocardiac myosin autoantibodies may be involved in the myocardial damage that occurs in this phase. In the final stage the vessels develop scarring, intimal thickening, calcification, and formation of thrombi. If death occurs as a result of this disease (rare), it is usually the result of aneurysm, coronary thrombosis, or severe scar formation and stenosis of the main coronary artery.

Clinical Manifestations. Clinical manifestations present in three phases: acute phase, subacute phase, and convalescent phase. In the *acute phase*, a sudden high fever (lasting over 5 days) that is unresponsive to antibiotics and antipyretics is followed by extreme irritability.

During the *subacute phase* (lasting approximately 25 days), the fever resolves, but the irritability persists along with other symptoms, such as anorexia, rash (exanthema) of the trunk and extremities with reddened palms and soles of the hands and feet, and subsequent desquamation (skin scales off) of the tips of the toes and fingers, peripheral edema of the hands and feet, cervical lymphadenopathy (usually unilateral), bilateral conjunctival infection without exudate, and changes in the oral mucous membranes (e.g., erythema, dryness and cracks or fissures of the lips, reddening or strawberry tongue).

In one third of all cases, children develop arthralgias and GI tract symptoms, typically lasting about 2 weeks. Joint involvement may persist for as long as 3 months. During this subacute phase, the person is at risk for cardiac involvement, especially the development of myocarditis, pericarditis, and arteritis that predisposes to the formation of coronary artery aneurysm in nearly 25% of cases not treated within 10 days of fever onset.

The *convalescent phase* occurs 6 to 8 weeks after onset of Kawasaki disease and is characterized by a resolution of all clinical signs and symptoms. However, during this phase the blood values have not returned to normal. At the end of the convalescent phase, all values return to normal and the child has usually regained his or her usual temperament, energy, and appetite.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Early recognition and prompt management of the acute syndrome are critical. Diagnosis is made on the basis of clinical manifestations and associated laboratory tests (there are no specific laboratory tests for Kawasaki disease). Echocardiograms are useful in providing a baseline and for monitoring myocardial and coronary artery status.

The introduction of high-dose IV immune globulin (IVIG) in combination with aspirin therapy to reduce fever and control inflammation and aneurysm formation has significantly reduced the prevalence of coronary artery abnormalities. The exact mechanism by which this treatment intervention reduces the vasculitis of acute Kawasaki syndrome has not been determined.

Prognosis is good for recovery with intervention, although serious cardiovascular problems (e.g., coronary thrombosis, aneurysm) may occur later in persons with cardiac sequelae. Giant aneurysms (diameter exceeding 8 mm) have the worst prognosis, since these are unlikely

to regress or resolve, with death common in this subgroup population. Occasionally, severe ischemic heart disease requires cardiac transplantation.¹⁹⁷

Thromboangiitis Obliterans (Buerger's Disease)

Overview and Pathogenesis. Thromboangiitis obliterans, also referred to as Buerger's disease, is a vasculitis (inflammatory and thrombotic process) affecting the peripheral blood vessels (both arteries and veins), primarily in the extremities. The cause is not known, but it is most often found in men younger than 40 years who smoke heavily, although the incidence in women is increasing. There has been some suggestion that unrecognized cocaine use may masquerade as Buerger's disease.²¹⁰

The pathogenesis of thromboangiitis obliterans is unknown; general inflammatory concepts apply. The inflammatory lesions of the peripheral blood vessels are accompanied by thrombus formation and vasospasm occluding and eventually obliterating (destroying) small and medium-sized vessels of the feet and hands.

Recent studies have linked elevated levels of homocysteine to Buerger's disease. Homocysteine has many potential effects: it limits the bioavailability of nitric oxide, impairs endothelium-dependent vasorelaxation, increases oxidative stress, stimulates smooth muscle cell proliferation, alters the elastic properties of vessel walls, and generates a prethrombotic state through the activation of factor V.

Clinical Manifestations. Clinical manifestations of pain and tenderness of the affected part are caused by occlusion of the arteries, reduced blood flow, and subsequent reduced oxygenation. The symptoms are episodic and segmental, meaning that the symptoms come and go intermittently over time and appear in different asymmetric anatomic locations. The plantar, tibial, and digital vessels are most commonly affected in the lower leg and foot. Intermittent claudication centered in the arch of the foot or the palm of the hand is often the first symptom.

When the hands are affected, the digital, palmar, and ulnar arteries are most commonly involved. Pain at rest occurs, with persistent ischemia of one or more digits. Other symptoms include edema, cold sensitivity, rubor (redness of the skin from dilated capillaries under the skin), cyanosis, and thin, shiny, hairless skin (trophic changes) from chronic ischemia. Paresthesias, diminished or absent posterior tibial and dorsalis pedis pulses, painful ischemic ulceration, and eventual gangrene may develop (see Fig. 12-29). Inflammatory superficial thrombophlebitis is common.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Arteriography may be used in the diagnosis, but definitive diagnosis of thromboangiitis obliterans is determined by histologic examination of the blood vessels (microabscesses in the vessel wall) in a leg amputated for gangrene. Given the new findings that cocaine use may present very much like Buerger's disease, laboratory screening for drug use may be appropriate in some cases.²¹⁰

Intervention should begin with cessation of smoking and avoidance of any environmental or secondhand smoke inhalation. All other treatment techniques are

aimed at improving circulation to the foot or hand, including pharmacologic intervention (e.g., vasodilators, pain relief) and physical or occupational therapy (see also Atherosclerosis: Medical Management).

Regional sympathetic ganglionectomy may produce vasodilation, ulcerations require wound care, and amputation (sometimes multiple) may be required when the individual is unable to quit smoking or when conservative care fails. With the recent finding of elevated levels of plasma homocysteine associated with Buerger's disease, screening for hyperhomocysteinemia and treatment of this condition have been recommended, especially to assess which clients may eventually require amputation.

Thromboangiitis is not life-threatening, but it can result in progressive disability from pain and loss of function secondary to amputation. Cessation of smoking is the key determinant in prognosis.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-18

Inflammatory Disorders

PREFERRED PRACTICE PATTERNS

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

5G: Impaired Motor Function and Sensory Integrity Associated with Acute or Chronic Polyneuropathies (peripheral neuropathy)

7A, 7D, 7E: (integumentary patterns) Associated with Buerger's disease

See also *Special Implications for the Therapist: Peripheral Vascular Disease*. Additional practice patterns may be appropriate if there is CNS involvement.

Peripheral neuropathy is a well-known and frequently early manifestation of many vasculitic syndromes. The pattern of neuropathic involvement depends on the extent and temporal progression of the vasculitic process that produces ischemia. A severe, burning dysesthetic pain in the involved area is present in 70% to 80% of all cases.

Other symptoms may include paresthesias and sensory deficit; severe proximal muscle weakness and muscular atrophy can occur secondary to the neuropathy. In the early phase, one nerve is affected and causes symptoms in one extremity (mononeuritis multiplex), but other nerves can become involved as the disorder progresses.

The therapist should watch for anyone with neuropathy who exhibits constitutional symptoms, such as fever, arthralgia, or skin involvement. This may herald a possible vasculitic syndrome and requires medical referral for accurate diagnosis. Early recognition of vasculitis can help prevent a poor outcome. With no treatment or with a poor outcome to intervention, CNS involvement (e.g., encephalopathy, ischemic and hemorrhagic stroke, cranial nerve palsy) can occur late in the course of vasculitis.

When corticosteroids (e.g., prednisone alone or sometimes in combination with other medications) are used (e.g., in the case of vasculitic neuropathy), the

therapist must be aware of the need for osteoporosis prevention and attend to the other potential side effects from the chronic use of these medications (see the section on Corticosteroids in Chapter 5).

Alternative methods of pain control may be offered in a rehabilitation setting, such as biofeedback, transcutaneous electrical nerve stimulation (TENS), and physiologic modulation (e.g., using a handheld temperature sensor to control autonomic nervous system function; see the section on Fibromyalgia in Chapter 7).

Vasculitis (Inflammatory Disease of Arteries and Veins)

The therapist's role in management of vasculitis may be primarily for relief of painful muscular and joint symptoms when present and in the prevention of functional loss in the case of neuropathies. For the client with thromboangiitis obliterans (Buerger's disease), exercise must be graded to avoid claudication and the client must be instructed in a home program for preventive skin care (see Box 12-14). Gangrene can occur as a result of prolonged ischemia from vessel obliteration; clients are typically treated for wound care and postoperatively after amputation. (See the section on Arteriosclerosis Obliterans below.)

Often a client with some other primary orthopedic or neurologic diagnosis has also been medically diagnosed with vasculitis (see the section on Rheumatoid Vasculitis in Chapter 5 and Special Implications for the Therapist boxes for clients with associated cardiovascular involvement, such as atherosclerosis, myocarditis, pericarditis, or aneurysm).

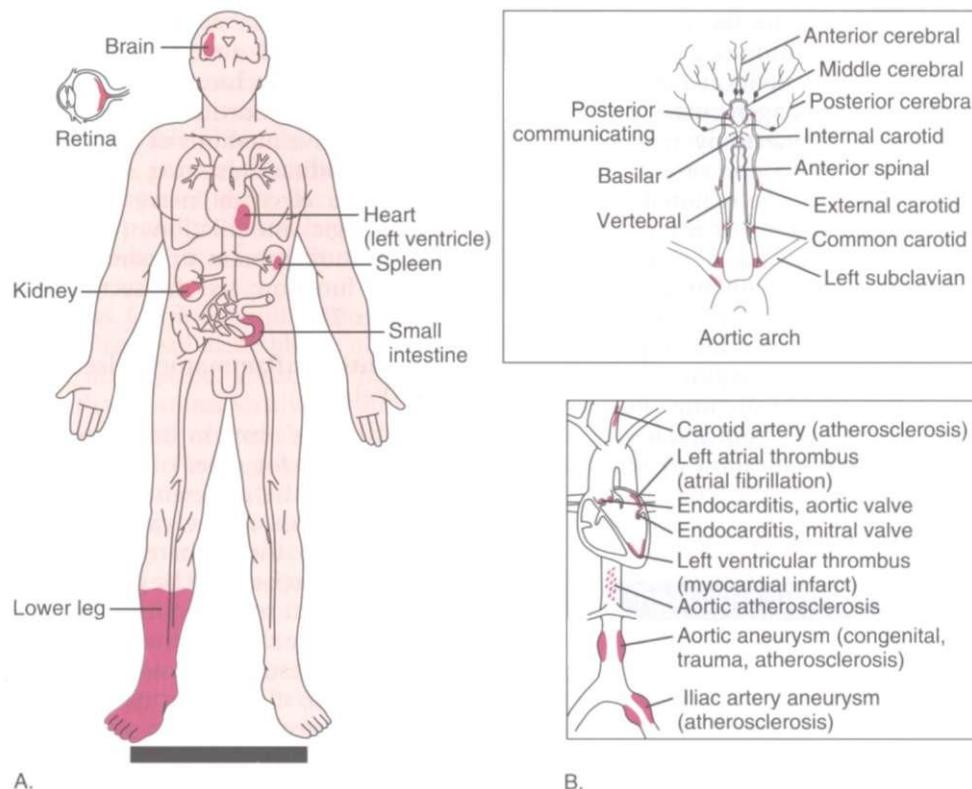
Arteritis

Early recognition and referral can prevent the serious complications associated with arteritis. Older adults who experience sudden or unexplained headaches, lingering flulike symptoms such as muscle aches (myalgia) and fatigue, persistent fever, unexplained weight loss, jaw pain when eating, or visual disturbances must be referred to their physicians. This is especially true for anyone with a previous diagnosis of PMR.

The use of corticosteroids can result in side effects such as osteoporosis and bone fractures, weight gain, diabetes, and high blood pressure (see complete discussion in Chapter 5). The client must be advised regarding an osteoporosis prevention program (see discussion in Chapter 24) and how to handle an increase in appetite. Remaining physically active and exercising are key components for both these issues.

Arterial Occlusive Diseases

Occlusive diseases of the blood vessels are a common cause of disability and usually occur as a result of atherosclerosis. Other causes of arterial occlusion include trauma, thrombus or embolism, vasculitis, vasomotor disorders such as Raynaud's disease or phenomenon and reflex sympathetic dystrophy (now called *complex regional pain syndrome*), arterial punctures, polycythemia, and

**Figure 12-28**

A, Common sites of infarction from arterial emboli. **B,** Sources of arterial emboli.

chronic mechanical irritation of the subclavian artery due to compression by a cervical rib. For each individual case, see the discussion of the underlying cause of the occlusion to understand etiologic and risk factors and pathogenesis.

Atherosclerotic occlusive disease can also affect other vessels throughout the body other than the cardiac blood vessels. For example, occlusive disease affecting the intestines results in acute intestinal ischemia or ischemic colitis (see Chapter 16), depending on the location of the occlusion.

Occlusive cerebrovascular disease (see Chapter 32) as a result of atherosclerosis accounts for many episodes of weakness, dizziness, blurred vision, or sudden cerebrovascular accident or stroke. Extracranial arterial ischemia (e.g., common carotid bifurcation, vertebral artery) accounts for over one half of these types of strokes.

Arterial Thrombosis and Embolism. Occlusive diseases may be complicated by arterial thrombosis and embolism (Fig. 12-28). Chronic, incomplete arterial obstruction usually results in the development of collateral vessels before complete occlusion threatens circulation to the extremity. Arterial embolism is generally a complication of ischemic or rheumatic heart disease, with or without MI.

Signs and symptoms of pain, numbness, coldness, tingling or changes in sensation, skin changes (pallor, mottling), weakness, and muscle spasm occur in the extremity distal to the block (Fig. 12-29). Treatment may include immediate or delayed embolectomy, anticoagulation therapy (e.g., heparin), and protection of the limb.

Thromboangiitis Obliterans (Buerger's Disease).

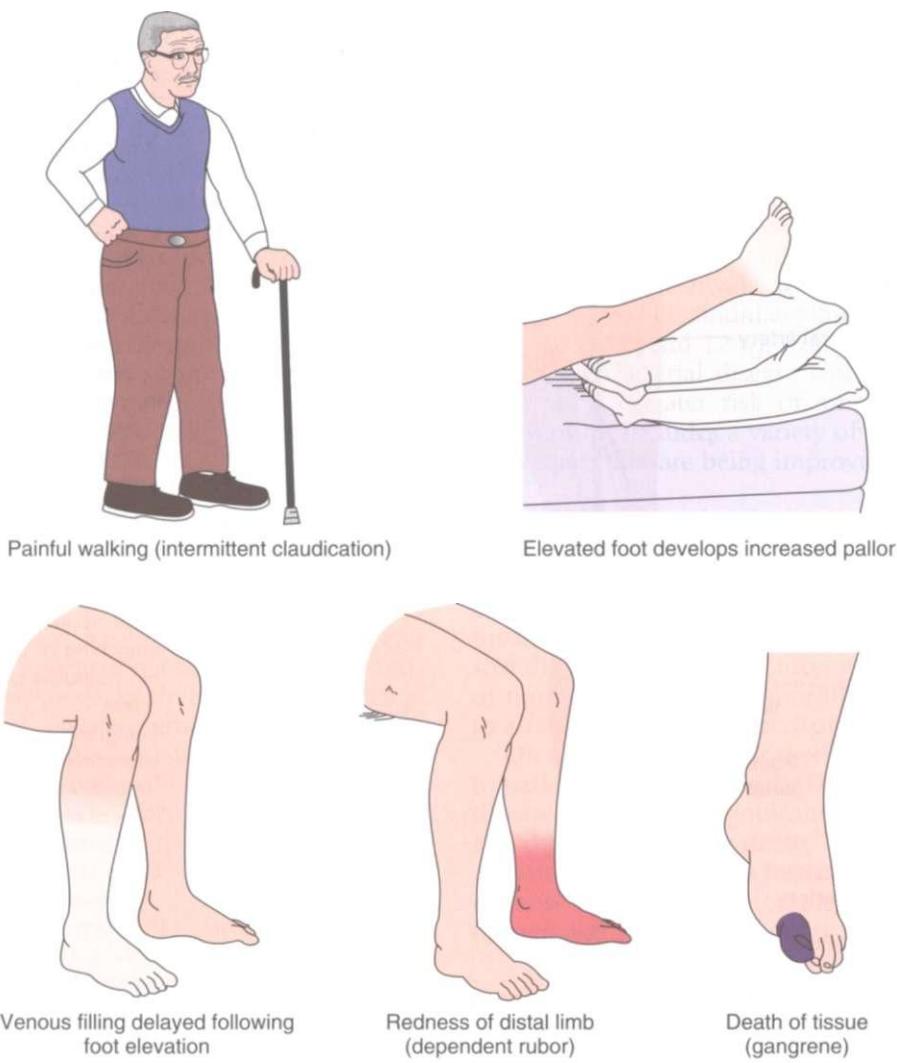
Thromboangiitis obliterans is discussed as a vasculitis in an earlier section (see Inflammatory Disorders) but is mentioned here as an occlusive disorder because the inflammatory lesions of the peripheral blood vessels are accompanied by thrombus formation and vasospasm, occluding blood vessels.

Arteriosclerosis Obliterans (Peripheral Arterial Disease)

Definition and Overview. Arteriosclerosis obliterans, defined as arteriosclerosis in which proliferation of the intima has caused complete obliteration of the lumen of the artery, is also known as atherosclerotic occlusive disease, chronic occlusive arterial disease, obliterative arteriosclerosis, and peripheral arterial disease (PAD). It is the most common arterial occlusive disease and accounts for about 95% of cases. It is a progressive disease that causes ischemic ulcers of the legs and feet and is most often seen in older clients, associated with diabetes mellitus.

Etiologic and Risk Factors. Atherosclerosis as the underlying cause of occlusive disease, with its known etiologic and associated risk factors, is discussed earlier in the chapter. PAD correlates most strongly with cigarette smoking and either diabetes or impaired glucose tolerance. Other risk factors include male gender, hypertension, low levels of HDL cholesterol, and high levels of triglycerides, apolipoprotein B, Lp(a), homocysteine, fibrinogen, and blood viscosity.

It has been reported that PAD is more prevalent in women than generally appreciated, but estimates vary

**Figure 12-29**

Signs and symptoms of arterial insufficiency.

greatly according to the diagnostic criteria applied. Prevalence and incidence rates do not differ significantly by gender, although incidence rates in women lag behind those in men in a pattern similar to that for CAD.²²⁹

Individuals with PAD are more likely to have CHD and cerebrovascular disease than those without PAD.²³⁰ There has been a debate as to usefulness of screening for PAD, and a conclusion has been made that targeted screening is likely to reduce heart attack, stroke, and death in patients with asymptomatic PAD.²³¹

Pathogenesis. See also Atherosclerosis: Pathogenesis. Since peripheral disease is one expression of atherosclerosis, understanding the pathogenesis of atherosclerosis is important. The arterial narrowing or obstruction that occurs as a result of the atherosclerotic process reduces blood flow to the limbs during exercise or at rest. Muscular reactivity is also adversely affected in PAD. Prostacyclin and nitric oxide usually activate vascular relaxation. In PAD, these relaxation factors are reduced and constrictive factors such as endothelin increase. This imbalance of vascular reactivity contributes to decreased blood flow.²⁹²

Clinical Manifestations. In peripheral vessels, claudication symptoms appear when the diameter of the vessel narrows by 50%. PAD affecting the lower extremities is primarily one of large and medium-sized arteries and most frequently involves branch points and bifurcations. Symptoms of arterial occlusive disease usually occur distal to the narrowing or obstruction. Acute ischemia may present with some or all of the classical symptoms, such as pain, pallor, paresthesia, paralysis, and pulselessness. However, arteries can become significantly blocked without symptoms developing, a phenomenon referred to as *silent ischemia*.

Even though silent ischemia is not associated with symptoms, it poses the same long-term sequelae and complications as overt ischemia and must be treated. It is strongly suspected when systolic blood pressure is lower at the ankle than at the arm (see further discussion of ankle/brachial index in this section).

Occlusive disease of the distal *aorta and iliac arteries* usually begins just proximal to the bifurcation of the common iliac arteries, causing changes in both lower extremities (Fig. 12-30; Table 12-19). Bilateral, progres-

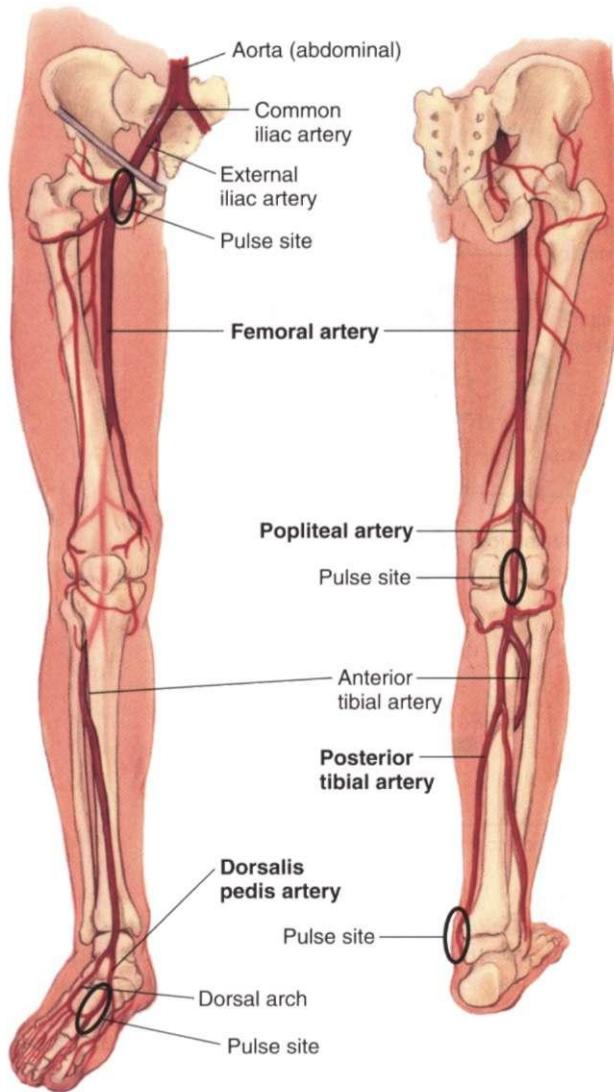


Figure 12-30

Arteries in the leg. The abdominal aorta branches (aortic bifurcation) into the right and left common iliac arteries. These arteries pass through the pelvic cavity and under the inguinal ligament to become the major arteries supplying the leg, called the femoral arteries. Each femoral artery travels down the thigh until, at the lower thigh, it courses posteriorly, where it becomes the popliteal artery. Below the knee, the popliteal artery divides into the anterior tibial artery and posterior tibial artery. The anterior tibial artery travels down the front of the leg onto the dorsum of the foot, where it becomes the dorsalis pedis artery. In the back of the leg, the posterior tibial artery travels down behind the malleolus and forms the plantar arteries in the foot. (From Jarvis C: *Physical examination and health assessment*, ed 5, Philadelphia, 2008, Saunders.)

sive, intermittent claudication (pain, ache, or cramp in the muscles causing limping) is almost always present in the calf muscles and is usually present in the gluteal and quadriceps muscles presenting as buttock, thigh, and calf pain.

The distance a person can walk before the onset of pain indicates the degree of circulatory inadequacy (e.g., two blocks or more is mild; one block is moderate; one half block or less is severe). The primary symptom may only be a sense of weakness or tiredness in these same

Table 12-19 Arterial Occlusive Disease

Site of Occlusion	Signs and Symptoms
Aortic bifurcation	Sensory and motor deficits Muscle weakness Numbness (loss of sensation) Paresthesias (burning, pricking) Paralysis
Iliac artery	Intermittent claudication (lower back, gluteal muscles, quadriceps, calves; relieved by rest) Cold, pale legs with decreased or absent peripheral pulses
Femoral and popliteal artery	Intermittent claudication (buttock, hip, thigh; relieved by rest) Diminished or absent femoral or distal pulses Impotence in males
Tibial and common peroneal artery	Intermittent claudication (calf, foot; may radiate) Leg pallor and coolness Dependent rubor Blanching of feet on elevation No palpable pulses in ankles and feet Gangrene Intermittent claudication (calves; feet occasionally) Pain at rest (severe disease); possibly relieved by dangling affected leg Same skin and temperature changes in lower leg and foot as described above Pedal pulses absent; popliteal pulses may be present

areas; both the pain and weakness or fatigue are relieved by rest.

Occlusive disease of the *femoral and popliteal arteries* usually occurs at the point at which the superficial femoral artery passes through the adductor magnus tendon into the popliteal space. Occlusion of these regions is also marked by intermittent claudication of the calf and foot that may radiate to the ipsilateral popliteal region and lower thigh. Although symptoms occur ipsilateral to the occlusion anywhere distal to the bifurcation of the aorta, most people have bilateral disease and therefore bilateral symptoms. There are definite changes of the affected lower leg and foot as listed in Table 12-19.

Occlusive disease of the *tibial and common peroneal arteries*, as well as the pedal vessels and small digital vessels, occurs slowly and progressively over months or years. The eventual outcome depends on the vessels that are occluded and the condition of the proximal and collateral vessels. Arterial ulcers may develop as a result of ischemia, usually located over a bony prominence on the toes or feet (e.g., metatarsal heads, heels, lateral malleoli). The skin is shiny and atrophic, and fissures and cracks are common.

Pain at rest indicates more severe involvement, which may mimic deep venous thrombosis (DVT), but relief from the occlusive disease can sometimes be obtained by

dangling the uncovered leg over the edge of the bed. This dependent position would increase symptoms of DVT, which is usually treated by leg elevation. Exercise may cause pedal pulses to disappear in some people. Sudden occlusion of the arteries, usually at the level of one of these smaller branches, results in gangrene. The necrotic tissue may become gangrenous and infected, requiring surgical intervention.

Occlusive arterial disease for the person with diabetes is further complicated by very slow healing, and healed areas may break down easily. In the case of diabetes mellitus, diabetic neuropathy with diminished or absent sensation of the toes or feet often occurs, predisposing the person to injury or pressure ulcers that may progress because of poor blood flow and subsequent loss of sensation (Table 12-20) (see the section on Diabetes in Chapter 11). Amputation rate in people with diabetes is markedly higher than for those individuals with PAD without diabetes mellitus.

MEDICAL MANAGEMENT

DIAGNOSIS. Diagnosis is based on client history and clinical examination. Diagnostic tools may include non-invasive vascular tests (e.g., ankle/brachial index, segmental limb pressures, pulse volume recordings, duplex ultrasonography) or, if invasive tests are required, arteriography with contrast or with MRI. An in-depth discussion of the diagnosis and intervention strategies for chronic arterial insufficiency of the lower extremities is available.⁸

PREVENTION AND TREATMENT. Prevention is the key to reducing the incidence of PVD caused by atherosclerosis. Risk factor reduction and lifestyle measures are the first steps, with smoking cessation (or not ever starting) as the single most effective prevention tool. A conservative approach to care that includes a program of dietary management to decrease cholesterol and fat, pain control, and daily physical training and exercise¹⁷ therapy to improve collateralization and function has been uniformly endorsed by experts in vascular disease.⁸ Careful attention must be given to preventive skin care (Box 12-14) to avoid even minor injuries, infections, or ulcerations.

Low-dose aspirin may be administered as an antiplatelet agent, and pentoxifylline may be used to improve capillary blood flow. Cilostazol (Pletal), with its unique combination of antiplatelet, vasodilatory, and antithrombotic effects, has been added to the pharmacologic agents for use with PAD. Although there are frequent minor adverse effects (e.g., headache, diarrhea, palpitations) the improved results with this agent (e.g., increased maximal walking distance 28% to 100%, increased pain-free walking distance 45% to 96%) may warrant its use over pentoxifylline.⁶⁴

Statins, used for the prevention of cardiovascular disease because of their antithrombotic properties, have been shown effective in reducing the risk of DVT and may become useful in the primary and secondary prevention of DVT.²⁷³

Surgical intervention (revascularization procedures such as bypass graft or angioplastic treatment; Fig. 12-31) is indicated if blood flow is compromised enough to

produce symptoms of ischemic pain at rest, if tissue death has occurred, or if claudication interferes with essential activities or work.⁶⁸ This decision is usually made after exercise therapy combined with risk factor modification has been unsuccessful in preventing this level of impairment and subsequent disability.

Cessation of smoking may be required by the physician before surgery is considered. Persons with localized occlusions of the aorta and iliac arteries less than 10 cm in length, with relatively normal vessels proximally and distally, are good candidates for angioplasty or stenting (see Figs. 12-3 and 12-5). Conversely, people with multisegmented arterial disease with more involved symptoms are at greater risk of amputation. Endovascular intervention includes a variety of catheter-based surgical techniques that are being improved by laser techniques.

PROGNOSIS. Arterial occlusive diseases are not life-threatening, but people with symptoms such as intermittent claudication often have a decreased quality of life because of mobility limitations. Symptoms of chronic arterial insufficiency progress slowly over time, so that progressive disability from pain, ulceration, gangrene, and loss of function or limbs is more likely to occur than death as a result of peripheral occlusive diseases.

On the other hand, because people with either asymptomatic or symptomatic PAD have widespread arterial disease, they have a significantly increased risk of stroke, MI, and cardiovascular death. Twenty percent of all individuals with PVD have a heart attack or stroke, and 30% of those have a 5-year mortality rate that climbs to 62% in men and 33% in women in 10 years.^{14,166}

SPECIAL IMPLICATIONS FOR THE THERAPIST

12-19

Arteriosclerosis Obliterans (Peripheral Arterial Disease)

See also Special Implications for the Therapist: Peripheral Vascular Disease.

Arterial Tests and Measures

Until recently, exercise testing using a 12-degree grade at a preset speed of 2.0 mph was the current practice to assess for claudication associated with PAD, but limited reproducibility and limited sensitivity to change in exercise performance were serious problems. Graded treadmill protocols have been developed to test people with PAD that give highly reproducible results and are able to evaluate change in exercise performance.

Two widely used graded protocols maintain a walking speed of 2.0 mph, one with grade increases of 3.5% every 3 minutes, the other with grade increases of 2.0% every 2 minutes. As the individual walks on the treadmill, time to pain and maximal walking time are recorded. All people limited by claudication are reproducibly brought to maximal levels of discomfort using either of these protocols.¹¹

Venous filling time provides a reasonable assessment of the general state of perfusion but requires

Continued.

Table 12-20 Comparison of Arterial, Venous, and Neuropathic Ulcers

	Arterial Ulcer	Venous Ulcer	Neuropathic (Diabetic) Ulcer
Etiology*	Arteriosclerosis obliterans Atheroembolism Large- or medium-vessel atherosclerosis Raynaud's disease Diabetes mellitus Collagen disease Vasculitis	Valvular incompetence History of deep venous thrombosis Venous insufficiency accompanied by hypertension Peripheral incompetence; varicose veins	Diabetes mellitus; combination of arterial disease and peripheral neuropathy Repetitive unrecognized trauma
Location	Anywhere on leg or dorsum of foot or toes Bone prominences (anterior tibial) Lateral malleolus Painful, especially with legs elevated Pulses poor quality or absent Intermittent claudication (exertional calf pain) Rest pain or nocturnal aching of foot or forefoot relieved by dependent dangling position Integumentary (trophic) changes Hair loss Thin, shiny skin Ischemia: pale, white skin color Areas of sluggish blood flow: Red-purple mottling Hypersensitivity to palpation History of minor nonhealing trauma	Medial aspect of distal one third of lower extremity Behind medial malleolus Can be very painful; venous insufficiency can cause aching pain; more comfortable with legs elevated Normal arterial pulses Edema Venous periwound (dark pigmentation) is called hemosiderin staining; leakage of hemosiderin is due to blood that cannot return because of vascular incompetence	Same areas in which arterial ulcers appear, especially toes Areas where peripheral neuropathy occurs [pressure points on plantar aspect of foot, toes, heels] Classic symmetric ascending stocking-glove distribution of sensory loss [begins in feet and ascends to knees, then symptoms begin in the hands] May not be painful because of loss of sensation (e.g., neuropathic ulcers are painless or insensitive when palpated) Some people experience unpleasant sensations (tingling or hypersensitivity to normally painless stimuli) Loss of vibratory sense and light touch Pulses may be present or diminished (arteries become calcified) Neuropathic foot is warm and dry Loss of vascular tone increases arteriovenous shunting and impairs blood flow necessary for wound healing; sepsis common Altered biomechanics and weight bearing
Clinical manifestations			
Wound appearance			
		Minimal exudate with dry necrosis Blanched wound base and periwound tissue	Superficial Highly exudative Red wound base Irregular edges
			Round, craterlike with elevated rim; diabetes hastens changes described in figure at left (arterial ulcer)
			Minimal drainage Frequently deep High infection rate

*Ulceration may also occur as a result of lymphatic disorders (see Chapter 13), skin cancer (see Chapter 9), metabolic abnormalities, and vasculitis (see Table 12-18).

Box 12-14**GUIDELINES FOR SKIN CARE AND PROTECTION****Temperature Protection**

- Nicotine causes vasoconstriction of the small vessels in the hands and feet; avoid all tobacco products.
- Recognize and avoid other triggers that cause vasoconstriction (e.g., emotional distress, caffeine, cold or cough remedies that contain a decongestant).
- Wear layers of clothing made of natural fibers, such as cotton, to draw moisture away from the skin; in cold weather, wear a hat and scarf, because heat is lost through the scalp; silk is a good insulator, consider it for socks and long underwear.
- Wear thick mittens, which are warmer than gloves, and socks purchased from an outdoor clothing or ski shop designed to wick moisture away while retaining body heat.
- Avoid air conditioning; wear warmer clothes, layer light clothing, or wear a sweater or jacket in air conditioning; be careful when going into an air-conditioned environment after being out in the heat or vice versa.
- Test water temperature before bathing or showering or have a member of the family test first; use other portion of the body to test if insensitivity exists in hand or foot.
- Use a heating pad, hot water bottle, or electric blanket to warm the sheets of your bed before getting into bed, but *do not* apply these directly to the skin and do not sleep with any electric device left on; if necessary wear light socks and mittens or gloves to bed. Do not soak hands or feet in hot water.
- Keep household temperatures at a constant, even, and comfortable level.
- Keep protective covering available at all times, even in the summer.
- Avoid contact with extremes of temperature, such as oven, dishwasher (hot dishes), refrigerator, or freezer; wear thick oven mitts whenever reaching into the oven. Keep mittens or warm gloves by the refrigerator and freezer to prevent symptoms when reaching into them.
- Wear rubber gloves whenever cleaning, washing dishes, or rinsing or peeling vegetables under water.
- Avoid holding ice, ice-cold fruit, hot or cold drinks, or frozen foods; wear protective gloves whenever making contact with any of these items.

Skin Care

- Take care of your skin and give your hands and feet extra care and protection; examine hands and feet daily; at the first sign of bruising, skin changes (e.g., cracking, calluses, blisters, redness), swelling, infection, or ulcer, immediately contact a member of your health care team (e.g., nurse, physical therapist, physician). If vision is impaired, have a family member or health care professional inspect your hands and feet.
- Circulation problems tend to create dry skin and delay healing; keep your skin clean and well moisturized; wash with a mild, creamy, or moisturizing liquid soap or gel; clean carefully between fingers and toes; *do not* soak them.
- Avoid perfumed lotions, and do not put lotion on sores or between toes.
- Observe carefully for any activities that might put pressure on your fingertips, such as using a manual typewriter, playing a musical instrument (e.g., guitar, piano), and doing crafts or needlework.

- Do not go barefoot indoors or outdoors; this includes getting up at night; avoid wearing open-toed shoes, pointy-toed shoes, high heels, or sandals; always wear absorbent socks or socks that wick perspiration away from skin; avoid nylon material (including pantyhose material); avoid stockings with seams or with mends; change socks or stockings daily.
- Make sure shoes provide good support without being too tight, avoid shoes that cause excessive foot perspiration, and alternate shoes throughout the week (i.e., do not wear the same shoes every day). Do not wear shoes without socks or stockings.
- Avoid hot tubs and prolonged baths; dry carefully between toes; water temperature should be between 90° and 95° F (32.2° and 35°C).
- Use heel protectors, sheepskin, and other protective devices whenever recommended.

Other Tips

- For Raynaud's disease or phenomenon, avoid situations that precipitate excitement, anxiety, or feelings of fear; teach yourself how to recognize early signs of these emotions and use relaxation techniques to reduce stress.
- For Raynaud's disease or phenomenon, when you have an attack, gently rewarm fingers or toes as soon as possible; place your hands in your armpits, wiggle fingers or toes, or move or walk around to improve circulation; if possible, run warm (*not* hot) water over the affected body part until normal color returns.
- Do not use razor blades; use electric razors.
- Avoid medications and substances (e.g., nicotine; caffeine in chocolate, tea, coffee, and soft drinks) that can cause blood vessels to narrow; discuss all medications with your physician.
- Maintain good circulation; do not stay in one position for more than 30 minutes; use breathing and stretching exercises whenever confined to a desk, chair, car, or bed for more than 30 minutes.
- Do not wear constricting or tight clothing, especially tight socks; avoid elastic around wrists or ankles.
- Do not wear jewelry, such as watches or bracelets, to bed at night.
- Leave a night light on in dark areas; turn on lights in dark areas and hallways.
- Do not sit with legs crossed because this can cause pressure on the nerves and blood vessels.
- Avoid sunburn.
- Do not scratch insect bites; do not scratch areas of itchy skin.
- Do not do bathroom surgery on corns or calluses; do not use chemical agents for the removal of corns or calluses; see your physician.

Care of Nails

- Use clippers, not scissors; *do not* use razor blades; cut toenails straight across, but file fingernails in a rounded fashion to the tips of your fingers.
- Take care of your nails; use cuticle softener or moisturizing cream or lotion around cuticles; push the cuticles back very gently with a cotton swab soaked in cuticle remover; *do not* push cuticles back with a sharp object and *do not* cut the cuticles with scissors or nail clippers.
- Use lamb's wool between overlapping toes.

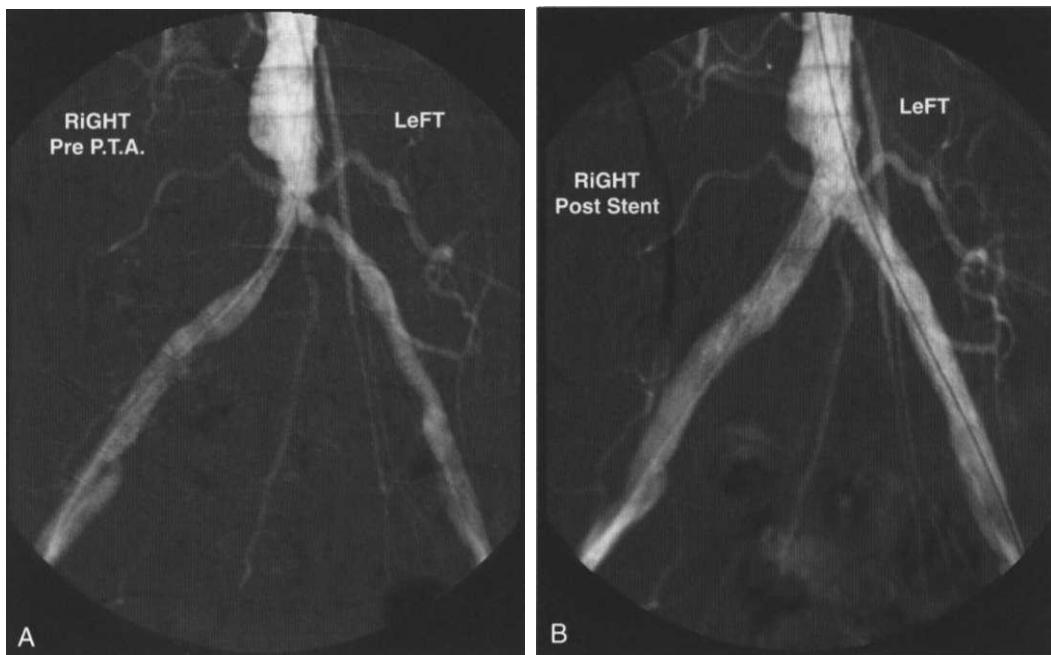


Figure 12-31

Percutaneous transluminal angioplasty (PTA) may be used in peripheral vascular disease. **A**, Significant narrowing of the aortic bifurcation and both common iliac arteries. The narrowing in both iliac arteries was successfully treated by angioplasty, and bilateral stents were inserted to maintain patency. **B**, The client had presented with bilateral calf claudication, which was relieved by this procedure. (From Forbes CD, Jackson WF: *Color atlas and text of clinical medicine*, ed 3, London, 2003, Mosby.)

patent venous valves to be a valid measure. Have the client assume a recumbent position and elevate the legs to facilitate venous drainage. When the veins have collapsed below the level of the skin, quickly bring the person to a sitting position with the legs hanging in a dependent position. The time necessary for the veins to fill to skin level is the venous filling time. A filling time greater than 25 seconds implies an increased risk of ulceration, infection, and poor wound healing.

The ankle/brachial index (ABI) (Box 12-15) is another measure of arterial perfusion at the level measured available to therapists for use in documenting the need for and benefit of a prescriptive exercise program. The ABI is a simple, inexpensive, and non-invasive tool that correlates well with angiographic disease severity and functional symptoms. It is well established as an independent predictor of cardiovascular morbidity and mortality.²²⁴

Blood pressures are measured both in the arm (brachial blood pressure) and the ankle with the client in a supine position for both measures. The ankle blood pressure may be auscultated using the dorsalis pedis pulse or posterior tibialis artery with the cuff placed above the ankle or using Doppler if available. The systolic ankle pressure is divided by the brachial systolic pressure.

With increasing degrees of arterial narrowing, there is a progressive fall in systolic blood pressure distal to the sites of involvement. If both pressures are measured with the person in the supine position and the vessels are unobstructed, the ratio of ankle to brachial pressures should be 1.0.^{2,2}

Box 12-15

ANKLE/BRACHIAL INDEX*

≥1.1	Suspicious for arterial calcification; blood vessels do not compress (e.g., diabetes)
1.0	Adequate blood supply; compression acceptable
<1.0	Inadequate blood supply; impaired wound healing; requires medical evaluation; prescriptive exercise beneficial
0.5-0.9	Indicates arterial occlusion; prescriptive exercise may be beneficial; delayed wound healing; light compression acceptable
<0.5	Severe arterial occlusive disease; may require surgical revascularization procedure; wound healing unlikely; rarely compress

*Values vary slightly from institution to institution and geographically from one area of the United States to another. Consider these values a general guideline and check the standard used at your current facility/location.

If flow to the lower extremity is decreased, the ratio will be less than 1.0. Based on two large population-based studies, the reference standard of ABI less than 0.90 at rest or less than 0.85 after exercise in adults older than 55 years indicates PAD.^{111,294} ABI measurements may be of limited value in anyone with diabetes, because calcification of the tibial and peroneal arteries may render them noncompressible.⁸

ABI can be measured before and after exercise to assess the dynamics of intermittent claudication. This can be accomplished by leaving the ankle pressure

cuffs in place during the exercise. Once the walk is completed or pain develops, the person rapidly assumes a supine position and the ankle pressures are measured.

At modest workloads, the healthy adult can maintain ankle systolic pressures at normal levels. If the exercise is strenuous, there may be a transient fall in systolic pressure that rapidly returns to baseline levels. In people with intermittent claudication, a different response is seen, even at low workload. If the person walks to the point of claudication, ankle systolic pressure falls precipitously, often to unrecordable levels, and will not return to baseline levels for several minutes.

It is not necessary (and may be misleading) to measure arm systolic pressure after exercise since this will increase by an amount related to the workload, and the most important variable is the extent to which ankle pressure falls and the time it takes to recover (i.e., the period of postexercise ischemia). In general, if ankle pressure falls by more than 20% of the baseline value and requires more than 3 minutes to recovery, the test result is considered abnormal.^{8,254}

Prescriptive Exercise

In prescribing an exercise program for someone with claudication secondary to occlusive disease, exercise tolerance must be determined. A training heart rate should be based on the exercise tolerance test, because persons with PVD frequently have CAD as well. Frequently, symptoms of claudication occur before training heart rate is reached, but the heart rate should be monitored and should not exceed the training heart rate, even in the absence of symptoms. Anginal chest pain is a red flag to decrease intensity.

A progressive conditioning program, including walking for fixed periods, is essential, even if the initial length of walking time is only 1 minute. Exercise can protect against atherothrombotic events by elevating t-PA³⁶⁵; improving peripheral circulation¹²¹; improving ambulatory function; increasing endothelial progenitor cells (cells that line the blood vessels capable of blood vessel repair),³⁰² endothelial-dependent dilation, and calf blood flow⁴⁹ and favorably altering cardiovascular risk factor profile (e.g., improved lipid profile, reduced blood pressure), an important element in the management of PAD.¹⁶³

The greatest improvement occurs with intermittent exercise to near-maximal pain progressing to a long-term (6 months or more) program of structured walking for at least 30 minutes three times weekly.¹²² The most effective program begins with brisk treadmill walking at a pace that is comfortable for the individual until claudication begins, followed by immediate rest and continued walking when the pain subsides.

The therapist can direct the client to progress quickly to levels of exercise at maximal tolerable pain in order to obtain optimal symptomatic benefit over time. This pattern is repeated starting with intervals as short as 1 to 5 minutes, alternating with rest periods of sufficient duration to eliminate pain (usually 2 to 10 minutes). Without complicating factors, the individual is usually

able to complete at least a 30- to 45-minute walk without pain or rest breaks within 6 to 8 weeks.

Claudication is influenced by the speed, incline, and surface of the walk and should be modified whenever possible to improve exercise tolerance. Impairment measures, functional measures, quality-of-life assessment, and specific walking parameters are outlined in detail elsewhere.²⁹² Supervised exercise and social support are recommended to improve both physical function and quality of life in people with PAD and intermittent claudication.²⁹¹

Altered gait pattern has been documented with PAD,¹²⁰ with less time spent in the swing phase of the gait cycle and more time in double stance. This ambulatory pattern favors greater gait stability at the expense of greater walking speed and can be improved with exercise rehabilitation. People with intermittent claudication associated with PAD are also functionally limited by dorsiflexion weakness, impairing their ability to perform tasks requiring distal lower extremity strength.²⁹⁷

After exercise, numbness in the foot as well as pain in the calf may occur. The foot may be cold and pale, which is an indication that the circulation has been diverted to the arteriolar bed of the leg muscles. Many people with claudication are already receiving β-blockers for angina or hypertension (see also Special Implications for the Therapist: Angina Pectoris and Special Implications for the Therapist: Hypertension earlier in this chapter).

The main factor limiting success of exercise therapy is lack of client motivation. For this reason, the most successful programs combine regular, supervised outpatient sessions with home exercise programs; regularity rather than intensity should be stressed.

Comorbid diseases, such as CAD or diabetes mellitus, and severity and location of arterial occlusive disease do not preclude successful response to prescriptive exercise. Unstable cardiopulmonary conditions require more careful consideration and collaboration with the health care team.⁸

Precautions

When arterial thrombosis or embolism is suspected, the affected limb must be protected by proper positioning below the horizontal plane and protective skin care provided. Heat or cold application and massage are contraindicated, and family members must also be notified of these restrictions. The home health therapist must be alert to the possibility of hot water bottles, heating pads, electric blankets, and hot foot soaks being used by the client without physician approval. This precaution is especially true for people with diabetes-associated peripheral neuropathies and for people with paraplegia.

Encourage the person with vascular disease to prevent becoming chilled by keeping the thermostat at home set at 70° to 72° F (21.1° to 22.2° C) and to avoid prolonged exposure to cold outdoors (e.g., by prewarming the car, dressing properly in layers, especially protecting hands and feet).

Continued.

In addition, many people with PVD and diabetes mellitus have peripheral sensory neuropathy and are at greater risk for skin breakdown on the foot from weight-bearing activities such as walking or running (see Box 12-14). These individuals should participate in alternate forms of exercise (e.g., bicycling, swimming/aquatics) even though these exercises may not improve walking ability as much as a structured walking program.²³⁰

Venous Diseases

Venous disease can be acute or chronic; acute venous disease includes thrombophlebitis, and chronic venous disease includes varicose vein formation and chronic venous insufficiency.

Venous Thrombosis and Pulmonary Embolus²³¹

Definition and Overview. Venous thrombosis is a partial occlusion (mural thrombus) or complete occlusion (occlusive thrombus) of a vein by a thrombus (clot) with secondary inflammatory reaction in the wall of the vein. A venous thrombus is an intravascular collection of platelets, erythrocytes, leukocytes, and fibrin, the end result of the clotting cascade with the potential to produce significant morbidity and mortality.²³²

There are two types of venous thrombosis: superficial (most commonly of the saphenous vein in the lower extremity; Fig. 12-32) and deep (usually of the femoral or iliac veins of the lower extremities and pelvis). Superficial venous thrombosis of the upper extremity can occur, although it is much less common and is usually seen in people with a systemic illness in the presence of an indwelling central venous catheter (e.g., used in the treatment of cancer), malignancy, or, less often, hemodialysis.²³³

In the lower extremities, superficial venous thrombus is usually the result of varicose veins, is self-limiting, and is not a serious condition. DVT of the lower extremity may be either a calf vein or proximal thrombosis (from the trifurcation of the popliteal vein caudally). Calf vein thrombosis is usually clinically silent and benign without complications, although silent calf vein thromboses can extend into more proximal veins (approximately 30% of the time).²³⁴ Proximal DVTs are much more likely to become PEs (see the discussion in Chapter 15).²³⁵

Incidence and Etiologic and Risk Factors. DVT is the third most common cardiovascular disease after acute coronary artery episodes and cerebrovascular accidents, affecting up to 2 million Americans annually.²³⁶

High-risk surgical candidates have a history of recent venous thromboembolism or have undergone extensive pelvic or abdominal surgery for advanced malignancy, CABG, renal transplantation, splenectomy, or major orthopedic surgery to the lower limbs (e.g., hip or knee arthroplasty, surgery for fractured hip, tibial osteotomy).

Approximately 30% to 60% of all people (women more than men) undergoing major general surgical procedures or having common pathologies such as cerebrovascular accidents develop clinical manifestations of DVT

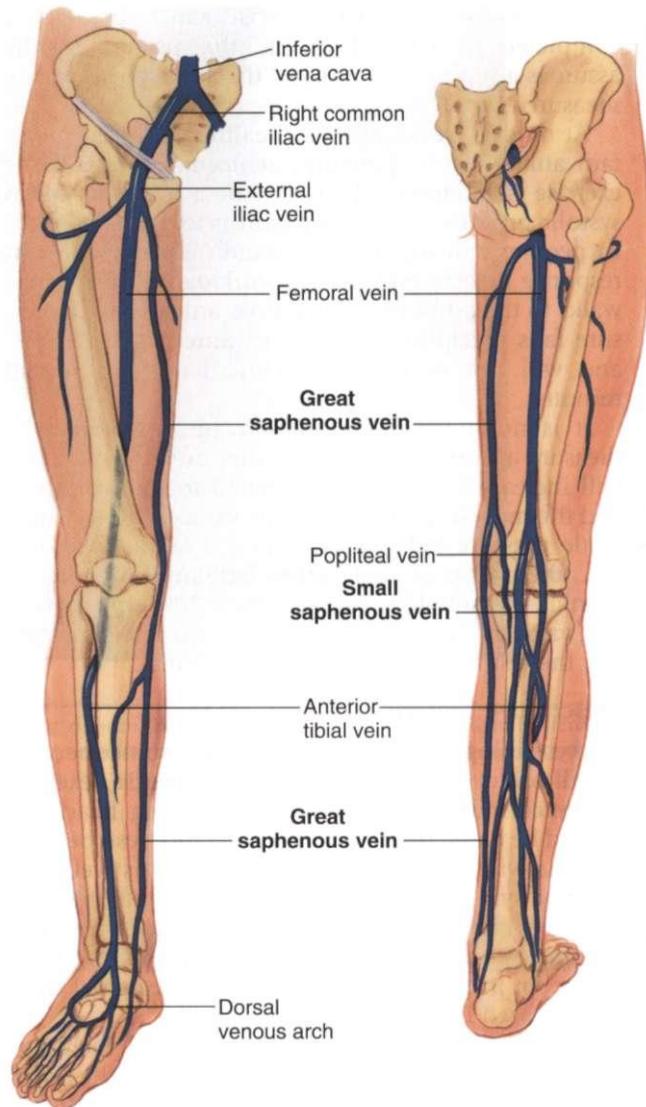
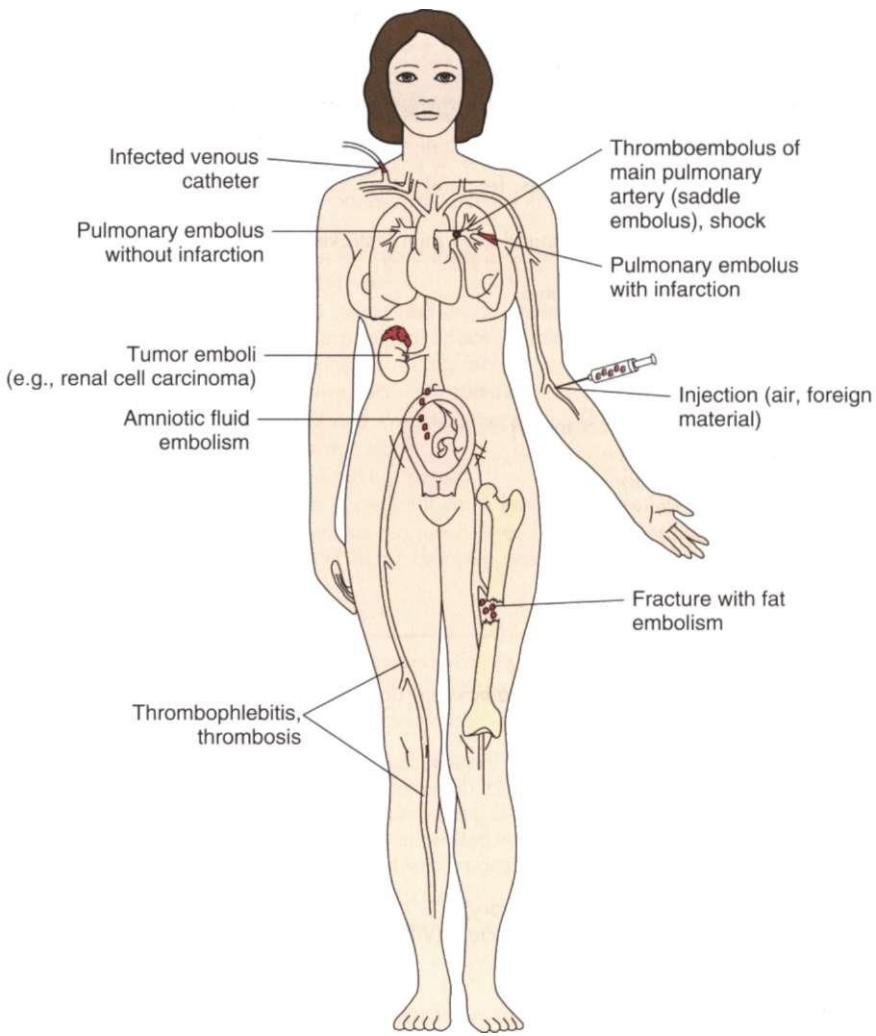


Figure 12-32

Veins in the leg. The legs have three types of veins: deep veins (femoral and popliteal) coursing alongside the deep arteries to conduct most of the venous return from the legs; superficial veins, the great and small saphenous veins; and perforators (not pictured), the connecting veins that join the two sets and route blood from the superficial into the deep veins. The great saphenous vein starts at the medial side of the dorsum of the foot and ascends in front of the medial malleolus, crossing the tibia obliquely and ascending along the medial side of the thigh. The small saphenous vein starts on the lateral side of the dorsum of the foot and ascends behind the lateral malleolus and up the back of the leg, where it joins the popliteal vein. (From Jarvis C: *Physical examination and health assessment*, ed 5, Philadelphia, 2008, Saunders.)

up to 4 weeks after the operation or incident^{237,238} (Fig. 12-33).

Fat embolism syndrome from fat thromboembolic phenomena is a well-known consequence of femoral total hip replacement arthroplasty. Intravasation of fat into the bloodstream during prosthetic implantation has been linked with postoperative confusion and cognitive decline. The risk of fat embolism syndrome is four times greater with simultaneous bilateral total knee or total hip

**Figure 12-33**

Sources and effects of venous emboli.

replacements.^{182,188,279} Changes have been made in the arthroplasty surgical technique that may result in a reduced incidence of this complication.

Brain microemboli from cardiac surgery with subsequent neurologic dysfunction has also been reported. The major source of the microemboli is lipid droplets of the patient's fat that drip into the blood in the surgical field. The lipid-laden blood is aspirated and returned to the patient via the cardiopulmonary bypass apparatus.³¹⁸

Substantial evidence indicates that the pathologic processes of venous (and arterial) thromboembolism involve both genetic and lifestyle influences. Scientific progress over the past decade has revealed a growing number of genetic factors present in more than 1% of the population that increase the relative risk of venous thrombosis between twofold and sevenfold. Several of these factors have been demonstrated to interact adversely with lifestyle influences, such as oral contraceptives and smoking.

Thrombus formation is usually attributed to venous stasis, hypercoagulability, or injury to the venous wall, although other risk factors may be present (Box 12-16). It is commonly held that at least two of these three conditions must be present for thrombi to form. What were previously considered to be idiopathic causes of throm-

bosis now have been identified as abnormalities associated with thrombophilia, such as elevated levels of coagulation factors VIII and XI, a particular prothrombin mutation, elevated levels of homocysteine, and a syndrome of activated protein C resistance.¹⁰⁶

Acquired resistance to the anticoagulant action of activated protein C may be associated with the increased risk of venous thrombosis associated with pregnancy, hormone replacement therapy, the use of oral contraceptives, and possibly even in vitro fertilization.⁸⁹

Pathogenesis. Any trauma to the endothelium of the vein wall exposes subendothelial tissues to platelets and clotting factors in the venous blood, initiating thrombosis. Platelets adhering to the vein wall attract the deposition of fibrin, leukocytes, and erythrocytes, forming a thrombus that may remain attached to the vessel wall. There are two types of thrombus: (1) mural thrombus where the thrombus is attached to the wall of the vein but does not occlude the vessel lumen, and (2) occlusive thrombus, which begins by attachment to the vessel wall and progresses to completely occlude the vessel lumen.

Both mural and occlusive thrombi may undergo one of the following forms of evolution or resolution: (1) lysis or dissolution or recanalization, in which the thrombus is dissolved away and blood flow through the veins

Box 12-16**RISK FACTORS FOR DEEP VENOUS* THROMBOSIS (DVT)****Immobility (Venous Stasis)**

- Prolonged bed rest (e.g., burns, fracture)
- Prolonged air travel
- Neurologic disorder (e.g., spinal cord injury, stroke)
- Cardiac failure
- Absence of ankle muscle pump

Trauma (Venous Damage)

- Varicose veins
- Surgery
- Local trauma (e.g., direct injury)
- Intravenous injections
- Fracture or dislocation
- Childbirth and delivery
- Sclerosing agents

Lifestyle

- Hormonal status
 - Oral contraceptive use
 - Hormone replacement therapy
 - Hormonal medications (e.g., tamoxifen, doxorubicin [Adriamycin])
 - Pregnancy
 - In vitro fertilization (?)
- Smoking

Hypercoagulation

- Hereditary thrombotic disorders
- Neoplasm (especially viscera, ovary)
- Increased levels of coagulation factors (VIII, XI)
- Prothrombin mutation
- Increased levels of homocysteine
- Activated protein C syndrome

Other

- Diabetes mellitus
- Genetic
- Obesity
- Previous deep vein thrombosis (DVT)
- Buerger's disease
- Age >60 years
- Idiopathic

*The terms venous and vein are used interchangeably in the literature and in this text.

returns; (2) organization, with the potential for removal of thrombus and vein; (3) extension, where the thrombus enlarges either proximally or distally; and (4) release of the thrombus to form a pulmonary embolism.

As an example of resolution, *mural thrombus* may undergo lysis, whereby the thrombus, in the presence of the body's own plasminogen, dissolves. These are usually clinically silent and represent the majority of DVTs, which often remain undiagnosed.

A mural thrombus may enlarge from the initial site of attachment, however. Clots can extend from the site of origin in both proximal and distal directions. The length of the vein can measure more than 1 m.

Occlusive thrombus may undergo restoration of the central venous canal and thereby recanalization may occur as a result of the healing process (this occurs in less than 10% of cases following DVT). Although blood flow is restored through the vein, the valves do not recover function, resulting in backflow of blood and other secondary functional and anatomic problems (e.g., stasis, venous stasis ulcers, risk of another DVT, pulmonary embolism from recurrent DVT).

Most occlusive thrombi heal by removal of the clot and the nonfunctional vein. Adherence of the thrombus to the vein often leads to phagocytic cell removal of both the clot and the vein followed by deposition of scar tissue (often leading to venous insufficiency under these circumstances, a condition sometimes referred to as post-thrombotic syndrome). Mobilization and compression stockings following acute DVT reduce the risk of post-thrombotic syndrome.^{258,269,319}

Finally, the thrombus may break off and become free floating as an *embolism*. The embolus travels through the progressively enlarging venous vessels and through the right side of the heart to the progressively narrowing pulmonary artery, where it may become lodged and occlude pulmonary circulation (PE).³²⁹ If a thrombus occludes a major vein (e.g., femoral vein, vena cava, axillary vein), the venous pressure and volume rise distally. However, if a thrombus occludes a deep small vein (e.g., tibial, popliteal), collateral vessels develop and relieve the increased venous pressure and volume. This is why the majority of PEs come from proximal DVTs.

Clinical Manifestations. In the early stages, approximately one half of the people with DVT are asymptomatic for any signs or symptoms in the affected extremity. The lower extremities are affected most often (more than 90%) but upper extremity venous thrombi can also develop, the latter usually presenting with edema of the involved extremity and pain.

When symptoms occur in the lower extremity, the client may report a dull ache, a tight feeling, or pain in the calf, often misdiagnosed as some other cause of leg pain. When symptoms are reported in the entire extremity, the condition is more extensive. In 80% of the cases with symptoms, the DVT is proximal, above the trifurcation of the popliteal vein. It is important to realize that proximal DVTs more often lead to severe consequences of DVT and at the time of diagnosis more than 50% of the affected individuals already have PEs.¹⁹

Signs are often absent; when present but taken alone, they may be variable and unreliable. Signs and symptoms include leg or calf swelling, pain or tenderness, dilation of superficial veins, and pitting edema. The skin of the leg and ankle on the affected side may be relatively warmer than on the unaffected side (check for temperature changes with the backs of the fingers or a skin thermometer). If venous obstruction is severe, the skin may be cyanotic.

Any of these symptoms can occur without DVT, possibly associated with other vascular, inflammatory, musculoskeletal, or lymphatic conditions that produce signs and symptoms similar to those of DVT.³²⁹

PEs (see Chapter 15), most often from the large, deep veins of the pelvis and legs, are the most devastating

complication of DVT and can occur without apparent warning. Signs and symptoms of PE are dependent on the size and location of the PE^{153,278} and may include the following¹²⁹:

- Possible sudden death
- Pleuritic chest pain
- Diffuse chest discomfort
- Tachypnea
- Tachycardia
- Hemoptysis
- Anxiety, restlessness, apprehension
- Dyspnea
- Persistent cough

UPPER EXTREMITY VENOUS THROMBOSIS. In the case of upper extremity superficial venous thrombosis, dull pain and local tenderness in the region of the involved vein may be accompanied by signs of superficial induration (firm or hard cord) and redness. Upper extremity superficial venous thrombosis is self-limiting and does not cause PE, since the blood flow to deeper veins is through small perforating venous channels. Iatrogenic superficial venous thrombosis is often secondary to prolonged IV catheter use.

Upper extremity DVT is an increasing problem with prolonged use of central venous lines. A recent study showed that over an 11-year period, 586 persons were diagnosed with upper extremity DVT. PE rates were significantly lower than what is usually found with lower extremity DVT and interestingly, location of the DVT did not seem to affect PE rate or mortality (again differing from lower extremity DVT).¹⁵²

MEDICAL MANAGEMENT

DIAGNOSIS. Utilization of the Wells and colleagues clinical decision rule (CDR) in anyone with suspected DVT clusters signs, symptoms, and risk factors and classifies the person's likelihood of having DVT as low, moderate, or high (Table 12-21). The CDR has been shown to be a reliable and valid tool for clinical assessment for predicting the risk of DVT in the lower extremity.^{21,22,185,350,352} The CDR has been specifically shown to be valid with orthopedic outpatients.²⁷⁷ A recent investigation revealed that physical therapists often underestimate the likelihood of DVT in high-risk individuals and frequently do not refer to a physician when they should.²⁷⁸

Low-risk clients as assessed by history and clinical examination with the CDR receive a D-dimer blood test (checking for fibrin breakdown products released from a thrombus). Moderate- to high-risk individuals receive Doppler duplex ultrasonography as a rapid screening procedure to detect thrombosis. Venous duplex ultrasonographic scanning has replaced contrast venography as the primary diagnostic test for DVT because it allows noninvasive visualization of the vein while simultaneously providing information on venous flow.

It is recognized that often other calf muscle strain or contusion may be difficult to differentiate from venous thrombosis; further diagnostic testing may be required to determine the correct diagnosis. Occasionally, a ruptured Baker cyst may produce unilateral pain and swelling in the calf. A history of arthritis in the knee of the same leg

Table 12-21 Wells' Clinical Decision Rule for Deep Venous Thrombosis (DVT)

Clinical Presentation	Score
Active cancer (within 6 mo of diagnosis or receiving palliative care)	1
Paralysis, paresis, or recent immobilization of lower extremity	1
Bedridden for more than 3 days or major surgery in the last 4 wk	1
Localized tenderness in the center of the posterior calf, the popliteal space, or along the femoral vein in the anterior thigh/groin	1
Entire lower extremity swelling	1
Unilateral calf swelling (more than 3 mm larger than unininvolved side)	1
Unilateral pitting edema	1
Collateral superficial veins (nonvaricose)	1
An alternative diagnosis is as likely (or more likely) than DVT (e.g., cellulitis, postoperative swelling, calf strain)	-2
Total points	

From Wells PS, Anderson DR, Bormanis J, et al: Value of assessment of pretest probability of deep-vein thrombosis in clinical management, *Lancet* 350:1795-1798, 1997. Used with permission.

Key:

- 2 to 0 low probability of DVT (3%)
- 1 to 2 Moderate probability of DVT (17%)
- 3 or more High probability of DVT (75%)

Medical consultation is advised in the presence of low probability; medical referral is required with moderate or high score.

and the disappearance of the popliteal cyst at the time symptoms develop are clues the physician can use to make the differentiation. Although Homans' sign was once used for differential diagnosis of acute DVT, it is no longer considered a sensitive or specific test for ruling in or out DVT.

PREVENTION. Primary prevention of DVT is important through the use of early mobilization for low-risk individuals and prophylactic use of anticoagulants (see Table 12-5) in people considered at moderate to high risk for DVT. While such interventions reduce the risk of DVT, it must be understood that even people receiving anticoagulant therapy may still develop DVTs. The highest incidence of DVT occurs with abdominal, thoracic, pelvic, hip, or knee surgical procedures; neurologic or other conditions leading to paresis or paralysis; and prolonged immobilization, cancer, and CHF.

Routine use of knee elastic stockings in all postoperative clients has been adopted in most hospitals, and many facilities use pneumatic pressure devices with on/off cycles applied for the first few hours after major surgery to mimic the calf pump. Once the person is able, ankle pumping is added, since this has been shown effective in increasing average peak venous velocity (flow) from the lower extremity with dorsiflexion of the ankle by over 200%, thereby reducing DVT while the person is immobilized.³⁵⁷

TREATMENT. The goals of DVT management are to prevent progression to pulmonary embolism, limit extension of the thrombus, limit damage to the vein, and prevent another clot from forming. Current therapy is to administer low-molecular-weight heparin (LMWH) followed by long-term oral anticoagulation (warfarin). Anticoagulation therapy for acute DVT prevents enlargement of the thrombus and allows for further attachment of the thrombus to the vessel wall, thereby reducing the likelihood of PE.³²⁹

Management of DVT changed dramatically with the introduction of LMWH used as a bridge to warfarin. This medication is more effective than the previously used unfractionated heparin, has fewer major bleeding complications, and does not require laboratory monitoring of coagulation test results to adjust medications, allowing for earlier hospital discharge or treatment at home. However, anticoagulation therapy does not effectively address the need to restore venous function in the thrombosed veins.

Formerly, symptomatic intervention included bed rest for 3 to 5 days to prevent emboli and pressure fluctuations in the venous system that occur with walking; elevation of the leg with the knee flexed until the edema and tenderness subsided; and continuous local application of heat to relieve venospasm, produce analgesia, and promote resolution of inflammation. Ambulation (while wearing elastic stockings) was permitted if local tenderness and swelling had resolved (usually after 7 days for calf thrombosis and 10 to 14 days for thigh or pelvic thrombosis).

Today, routine practice is to authorize ambulation in all cases of DVT after adequate anticoagulation by LMWH or unfractionated heparin has been administered if local symptoms and general condition permit. The concern in an acute care or rehabilitation setting is the increased risk of PE in clients who are aggressively mobilized too soon after a diagnosis of a DVT and before adequate anticoagulation has been administered. Bed rest (up to 24 hours) may be advised before returning a person with acute DVT to ambulation and physical therapy.

A large cohort prospective study to assess the risk of PE with early mobilization has been recommended.¹⁸ Several randomized controlled trials and large registry trials have shown no increased risk of PE in the mobilized (ambulation with compression) group as compared to the immobilized (strictly bed rest) group.^{40,171,255-257,334}

Elastic stockings must be worn whenever the person is ambulating or in the upright position. The standard of care for DVT is moving toward the following protocol: inject with LMWH, discharge to home with additional doses the client can use to inject at home over the next week while taking warfarin. Return for follow-up in 1 week with evaluation of INR. Clients are advised to remain active but avoid any straining maneuvers.

For cases of massive DVT, thrombolysis, thrombectomy, and embolectomy (often performed in an interventional angiography laboratory) are being used with increasing skill and improved outcomes. New research is leading to the next generation of antithrombotic compounds, such as direct coagulation factor inhibitors; tissue factor pathway inhibitors; the use of statins²⁷³; and gene therapy.

Gene therapy for antithrombotic strategies can involve a number of different approaches, such as inhibition of coagulation factors, overexpression of anticoagulant factors, or modulation of endothelial biology to make thrombus formation or propagation unfavorable.

Other investigators are looking at the systemic administration of recombinant tissue factor pathway inhibitor to decrease intimal hyperplasia after vascular injury and to suppress systemic mechanisms of blood coagulation and thrombosis.³⁶⁹

PROGNOSIS. DVTs that are not diagnosed can lead to life-threatening consequences, such as PE. With appropriate intervention and in the absence of complications, a return to normal health and activity can be expected within 1 to 3 weeks for the person with a calf DVT and within 6 weeks for the person with thigh or pelvic DVT.

Prognosis depends on the size of the vessel involved, the presence of collateral circulation, and the underlying cause of the thrombosis (e.g., spinal cord injury, stroke, or neoplasm may prevent return to former health). Recurrence occurs in 5% of DVT cases and 1% of PE cases and may be related to risk factors listed in Box 12-16 or too short a time on anticoagulants.

It remains unknown whether anticoagulant therapy should be extended for longer periods (more than 3 months), if lower intensity should be recommended during this extension (i.e., with a target INR of less than 2.0), or if the benefits of extended anticoagulant therapy outweigh the risk of bleeding complications.

A potential long-term complication of DVT is venous stasis or insufficiency (postthrombotic syndrome) when permanent damage to the vein has occurred; see the section on Chronic Venous Insufficiency in this chapter. Between 25% and 30% of people who had DVT treated with anticoagulants will develop some form of postthrombotic syndrome in the first 10 years following DVT.

The National Institutes of Health are investigating the combined use of anticoagulants and thrombolytics to preserve the patency of the veins, thereby reducing the frequency of postthrombotic syndromes compared to anticoagulation therapy alone. Previously, the prohibitive cost, need for hospitalization, and procedural complications associated with thrombolytic therapy have prevented their use in proximal DVT.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-20

Venous Thrombosis and Pulmonary Embolism

PREFERRED PRACTICE PATTERNS

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

7A: Primary Prevention/Risk Reduction for Integumentary Disorders

Risk Assessment

Populations at risk (see Box 12-16), especially postoperative, postpartum, and immobilized clients, should

be identified by the medical staff and observed carefully. Risk factor assessment scales (e.g., Autar DVT scale) for use in a therapy practice are available.^{28,29,329} Using seven risk categories (i.e., increasing age, BMI, immobility, special DVT risk, trauma, surgery, high-risk disease), the therapist can accurately predict and categorize each person's risk for venous thromboembolic disease as no risk (less than 10%), low risk (10%), moderate risk (11% to 40%), or high risk (more than 41%).³²⁹

The person at risk for DVT secondary to fracture and subsequent immobility involving a lower extremity cast should be carefully evaluated when the cast is removed. Normally, calf muscle atrophy is easily observed when the cast is removed. Normal calf size (less than 1-cm difference between left and right) without atrophy on cast removal may signal swelling associated with DVT.

For the client with diagnosed thrombophlebitis, the therapist should monitor and report any signs of PE, such as chest pain, hemoptysis, cough, diaphoresis, dyspnea, and apprehension. Clients with a history of DVT may develop chronic venous insufficiency even years later and therefore must be monitored periodically for life.

Anyone receiving anticoagulant therapy (e.g., warfarin [Coumadin]) must be monitored for manifestations of bleeding, as evidenced by blood in the urine, in the stool, or along the gums or teeth; subcutaneous bruising; or back, pelvic, or flank pain. The presence of any of these signs or symptoms must be reported to the physician immediately.

The risk for bleeding is increased with alcohol use, especially if there is concomitant liver disease, since alcohol also can potentiate warfarin. Many herbs have natural anticoagulant effects that can potentiate the effect of warfarin, and others can counteract its effect. Ginkgo biloba, garlic, dong quai, dan shen, and ginseng (herbs commonly ingested in supplemental form) should not be used or taken at the same time as warfarin. Anyone using these products should be encouraged to discuss medication dosage with the prescribing physician. Eating large quantities of vitamin K-rich foods can also interfere with the drug's anticoagulant effects, requiring careful monitoring of food intake while on warfarin.

Bleeding under the skin and easy bruising in response to the slightest trauma can occur when platelet production is altered. This condition necessitates extreme care in the therapy setting, especially any intervention requiring soft tissue mobilization, manual therapy, or the use of any equipment, including any modalities and weight-training devices. (See the section on Platelets in Chapter 40; see also Tables 40-8, 40-9, and 40-12.) Rarely, skin necrosis associated with the use of warfarin occurs, presenting as large hemorrhagic blisters on the breasts, buttocks, thighs, and penis requiring wound management.

Prevention and Intervention

Prevention is the key to treatment of thrombophlebitis, both preventing thrombus formation and prevent-

ing thrombi from becoming emboli. Preventive therapy can be tailored to the individual's level of risk and may include active and passive range-of-motion exercise, early ambulation for brief but regular periods whenever possible, coughing and deep-breathing exercises, and proper positioning.

The person at risk must be taught the importance of avoiding one position for prolonged periods and avoiding pillows under the legs postoperatively to facilitate venous return. At the same time, elevation of the legs just above the level of the heart aids blood flow by gravitational force and prevents venous stasis as a contributing factor to the formation of new thrombi.

Placing the foot of the bed in Trendelenburg's position (6-inch elevation with slight knee bend to prevent popliteal pressure) decreases venous pressure and helps relieve pain and edema. Prolonged sitting in a chair in the early postoperative period should be avoided.

After thrombosis of a deep calf vein, elastic support hose should be worn for at least 6 to 8 weeks or longer if risk assessment is moderate or high. Helping the client find easier ways to put the hose on and explaining the purpose may increase compliance in using the hose consistently and correctly.

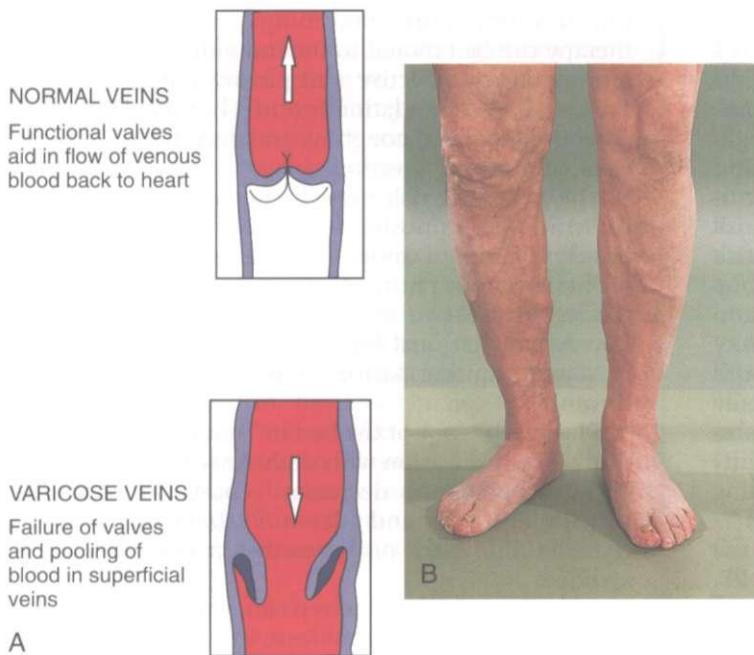
Support pantyhose may be an acceptable alternative for some people who have trouble putting on the compressive stockings or who live in very hot climates. The hypothesis for the use of compressive or elastic stockings is that the compressive force applied by the stocking causes the vessel wall to become applied to the thrombus, thereby keeping the thrombus in its location and preventing movement inside the blood vessel. Without the external compressive force of the stocking, once the person stands, increased hydrostatic pressure causes venous distention and permits the thrombus to become free floating inside the vessel.³²⁹

Varicose Veins

Definition and Incidence. Varicose veins are an abnormal dilation of veins, usually the saphenous veins of the lower extremities, leading to tortuosity (twisting and turning) of the vessel, incompetence of the valves, and a propensity to thrombosis. Women are affected with leg varicosities more often than men (secondary to pregnancy) until age 70 years, when the gender difference disappears. Forty-one percent of women ages 40 to 50 years and 72% of women ages 60 to 70 years have varicose veins.¹⁴²

This condition most often develops between the ages of 30 and 50 years for all persons. A separate but similar condition called *spider veins* or *telangiectasia* (broken capillaries) results in fine-lined networks of red, blue, or purple veins, usually on the thighs, calves, and ankles. The veins may form patterns resembling a sunburst, a spider web (see Fig. 10-3), or a tree with branches but can also appear as short, unconnected, or parallel lines.

Etiologic and Risk Factors. Varicose veins may be an inherited trait, but it is unclear whether the valvular

**Figure 12-34**

A, Diagrams of normal (top) and varicose (bottom) veins. **B**, Person with varicose veins. (A, from O'Toole M, ed: Miller-Keane encyclopedia and dictionary of medicine, nursing, and allied health, ed 6, Philadelphia, 1997, Saunders, p 1702. B, from Forbes CD, Jackson WF: Color atlas and text of clinical medicine, ed 3, London, 2003, Mosby.)

incompetence is secondary to defective valves in the saphenous veins or to a fundamental weakness of the walls of the vein leading to dilation of the vessel.

Periods of high venous pressure associated with heavy lifting or prolonged sitting or standing are risk factors. Hormonal changes (e.g., pregnancy, menopause, hormonal therapy) often contribute to the development of this condition by relaxing the vein walls.

Other risk factors include pressure associated with pregnancy or obesity, heart failure, hemorrhoids, constipation, esophageal varices, and hepatic cirrhosis. Risk factors for spider veins are similar (age, hormones, familial predisposition) but also include local injury (past or present).

Pathogenesis. Blood returning to the heart from the legs must flow upward through the veins, against the pull of gravity. This blood is milked upward, principally by the massaging action of the muscles against the veins. To prevent the blood from flowing backward, the veins contain one-way valves located at intervals, which operate in pairs by closing to stop the reverse movement of the blood.

The vessels most commonly affected by varicosities are located just beneath the skin superficial to the deep fascia and function without the kind of support deep veins of the legs receive from surrounding muscles. As the one-way valves become incompetent or the veins become more elastic, the veins engorge with stagnant blood and become pooled.

Any condition accompanied by pressure changes places a strain on these veins, and the lack of pumping action of the lower leg muscles causes blood to pool. Other sites involved include the hemorrhoidal plexus of the rectum and anal canal (either inside or outside the anal sphincter), submucosal veins of the distal esophagus, and the scrotum (varicocele).

The weight of the blood continually pressing downward against the closed venous valves causes the veins to distend and eventually lose their elasticity. When several valves lose their ability to function properly, the blood collects in the veins, causing the veins to become swollen and distended. During pregnancy, the uterus may press against the veins coming from the lower extremities and prevent the free flow of returning blood. More force is required to push the blood through the veins, and the increased back-pressure can result in varicose veins.

Clinical Manifestations. The clinical picture is not directly correlated with the severity of the varicosities; extensive varicose veins may be asymptomatic, but minimal varicosities may result in multiple symptoms. The development of varicose veins is usually gradual; the most common symptom reported is a dull, aching heaviness, tension, or feeling of fatigue brought on by periods of standing. Cramps of the lower legs may occur, especially at night, and elevation of the legs often provides relief. Itching from an associated dermatitis may also occur above the ankle.

The most visible sign of varicosities is the dilated, tortuous, elongated veins beneath the skin, which are usually readily visible when the person is standing (Fig. 12-34). Varicosities of long duration may be accompanied by secondary tissue changes, such as a brownish pigmentation of the skin and a thinning of the skin above the ankle. Swelling may also occur around the ankles.

Untreated, the veins become thick and hard to the touch; impaired circulation and skin changes may lead to ulcers of the lower legs, especially around the ankles (see Table 12-20). (See also the section on Esophageal Varices in Chapter 16.) One of the most important distinctions between varicose veins and spider veins is that, in some cases, varicose veins can result in thromboses (blood clots) and phlebitis (inflammation of the vein) or

venous insufficiency ulcers. Spider veins are merely a cosmetic issue with no adverse effects.

MEDICAL MANAGEMENT

DIAGNOSIS. The physician must distinguish between the symptoms of arteriosclerotic PVD, such as intermittent claudication and coldness of the feet, and symptoms of venous disease, because occlusive arterial disease usually contraindicates the operative management of varicosities below the knee. When the two conditions coexist, the reduced blood flow caused by the atherosclerosis may even improve the varicosities by reducing blood flow through the veins.

Visual inspection and palpation identify varicose veins of the legs, and Doppler ultrasonography or the duplex scanner is useful in detecting the location of incompetent valves. Endoscopy or radiographic diagnosis identifies esophageal varices, rectal examination or proctoscopy is used to diagnose hemorrhoids, and palpation identifies varicocele (scrotal swelling).

TREATMENT. Treatment of mild varicose veins is conservative, consisting of periodic daily rest periods with feet elevated slightly above the heart. Client education as to the importance of promoting circulation is stressed, including instructions to make frequent changes in posture, a daily exercise program, and the appropriate use of properly fitting elastic stockings.

When varicosities have progressed past the stage at which conservative care is helpful, surgical intervention and compression sclerotherapy may be considered. In the past, surgical treatment of varicose veins consisted of removing the varicosities and the incompetent perforating veins (ligation and stripping), a procedure sometimes referred to as *stripping the veins or miniphlebectomy*.

Other procedures for varicose veins have been developed, including radiofrequency (radio waves used to seal off the vein), sclerotherapy (injections of a hardening, or sclerosing, solution; over several months' time, the injected veins atrophy and blood is channeled into other veins), and laser therapy (noninvasive use of near-infrared wavelengths). Ligation and stripping of the greater saphenous vein prevent its use as a source for future CABGs, motivating researchers to develop effective intervention techniques that salvage large veins.

Oral dietary supplementation has been adopted by some individuals as an addition to traditional management of varicose veins. The loss of vascular integrity associated with the pathogenesis of both hemorrhoids and varicose veins may be aided by several botanical extracts shown to improve microcirculation, capillary flow, and vascular tone while strengthening the connective tissue of the perivascular substrate.²⁰⁴

PROGNOSIS. Good results with relief of symptoms are usually possible in the majority of cases. Early conservative care for varicose veins during initial stages may help prevent the condition from worsening, but advanced disease may not be prevented from recurring, even with surgical intervention or sclerotherapy. Although surgery for varicose veins can improve appearance, it may not reduce the physical discomfort, suggesting that most

lower limb symptoms may have a nonvenous cause. A high mortality is associated with ruptured, bleeding esophageal varices (see Chapter 16).

SPECIAL IMPLICATIONS FOR THE THERAPIST

12-21

Varicose Veins

PREFERRED PRACTICE PATTERNS

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

7A: Primary Prevention /Risk Reduction for Integumentary Disorders (other Integumentary patterns may apply depending on progression of disease or chronic complications)

The therapist can be very instrumental in developing prescriptive exercise and preventive measures for anyone at risk for, or already diagnosed with, varicose veins. Since excessive sitting or standing contributes to this condition, the therapist can individualize a program to help the person avoid static postures and utilize quick stretch or movement breaks coordinated with deep-breathing exercises.

Over-the-counter pantyhose should be replaced with special compressive hose that do not constrict behind the knee, upper leg, waist, or groin. These should be worn as much as possible during the daytime hours (including during exercise for some people) but may be removed at night. After exercise and at the end of the day, instruct the individual to elevate the legs in a supported position above the level of the heart for 10 to 15 minutes.

Encourage the person to practice good breathing techniques during this time. Aerobic exercise, strength training, or resistive exercises are encouraged, but high-impact activities, such as jogging or step aerobics, should be avoided. Brisk walking, cycling, cross-country skiing or Nordic track, rowing, and swimming are all good alternatives to high-impact activities.

Chronic Venous Insufficiency

Definition and Incidence. Chronic venous insufficiency (CVI), also known as postphlebitic syndrome and venous stasis, is defined as inadequate venous return over a long period of time. This condition follows most severe cases of DVT, although it is possible to develop CVI without prior episodes of DVT. CVI may also occur as a result of leg trauma, varicose veins, and neoplastic obstruction of the pelvic veins. The long-term sequelae of CVI may be chronic leg ulcers, accounting for the majority of vascular ulceration; incidence is expected to continue rising with the aging of America.²³⁵

Etiologic Factors and Pathogenesis. CVI occurs when damaged or destroyed valves in the veins result in decreased venous return, thereby increasing venous pressure and producing venous stasis. Without adequate valve function and in the absence of the calf muscle pump, blood flows in the veins bidirectionally, causing high ambulatory venous pressures in the calf veins (venous

hypertension). Superficial veins and capillaries dilate in response to the venous hypertension. Red blood cells, proteins, and fluids leak out of the capillaries into interstitial spaces, producing edema and the reddish brown pigmentation characteristic of CVI.

Chronic pooling of blood in the veins of the lower extremities prevents adequate cellular oxygenation and removal of waste products. Any trauma, especially pressure, further lowers the oxygen supply by reducing blood flow into the area. Cell death occurs, and necrotic tissue develops into venous stasis ulcers. The cycle of reduced oxygenation, necrosis, and ulceration prevents damaged tissue from obtaining necessary nutrients, causing delayed healing and persistent ulceration. Poor circulation impairs immune and inflammatory responses, leaving venous stasis ulcers susceptible to infection.

Other contributing factors may include poor nutrition, immobility, and local trauma (past or present). A previous history of burns requiring skin grafts predisposes the individual to venous insufficiency. The area of the graft usually lacks superficial veins, properly functioning capillaries, or both, resulting in blood pooling in these areas. As a result previously burned areas and skin grafts in the lower extremity are susceptible to vascular ulceration.

Clinical Manifestations. CVI is characterized by progressive edema of the leg; thickening, coarsening, and brownish pigmentation of skin around the ankles; and venous stasis ulceration (see Table 12-20). Venous insufficiency ulcers constitute approximately 80% of all lower extremity ulcers, occurring most often above the medial malleolus where venous hypertension is greatest.

These ulcers characteristically are shallow wounds with a white creamy to fibrous slough over a base of good granulation tissue. They can be very painful with a moderate to large amount of drainage. The wounds typically have irregular borders and are partial to full thickness, often with signs of reepithelialization (e.g., pink or red granulation base). Frequently, moderate to severe edema is present in the limb; in longstanding cases, this edema becomes hardened to a dense, woody texture. The skin of the involved extremity is usually thin, shiny, dry, and cyanotic. Dermatitis and cellulitis may develop later in this condition.

MEDICAL MANAGEMENT

The physician will differentiate between CVI and other causes of edema and ulceration of the lower extremities using client history, clinical examination, and diagnostic tests to rule out or confirm superimposed acute phlebitis. Arterial and venous insufficiency may coexist in the same person.

Treatment goals and techniques are as for varicose veins (increase in venous return, reduction of edema). Conventional methods of compression and rest and elevation (e.g., more frequent periods of leg elevation above the level of the heart are encouraged throughout the day with the foot of the bed elevated 6 inches at night) have been augmented by surgical intervention.

Rapid progress in endovascular procedures with angioplasty and stenting has made it possible for the development of techniques to relieve obstruction and repair reflux in the deep veins. Venous stasis ulcers require

ongoing treatment, usually involving the therapist (e.g., primary intervention for edema reduction and topical ulcer and wound care). More detailed information is available.^{155,181,321,322} Researchers are developing bioengineered skin, a living human dermal replacement for the management of venous ulcers. See the section on Skin Transplantation in Chapter 21.

The prognosis is poor for resolution of CVI, with chronic venous stasis ulcers causing loss of function and progressive disability. Recurrent episodes of acute thrombophlebitis may occur, and noncompliance with the treatment program is common.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-22

Chronic Venous Insufficiency

PREFERRED PRACTICE PATTERNS

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

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

7B: Impaired Integumentary Integrity Associated with Superficial Skin Involvement (other Integumentary patterns may apply depending on progression of the condition)

The therapist can be very instrumental in providing clients with venous insufficiency with education and prevention to avoid complications that can occur with vascular ulceration and chronic wounds. Formulating an exercise prescription; collaborating with a nutritionist; and understanding the underlying etiology, hemodynamics, comorbidities, and principles of tissue repair are essential in developing a plan of care.

Compression therapy (e.g., bandages, stockings, pumps) is the gold standard for treatment of venous insufficiency, especially when venous leg ulcers are present. The goal is to promote venous return from peripheral veins to central circulation. The therapist may also use layered gradient compression wraps (see Fig. 10-9, C). The presence of CHF is considered a precaution to the use of external compression and requires close collaboration between the physician and therapist.

Before initiating compression therapy the ABI should be measured. ABI is determined using a non-invasive arterial Doppler study to assess the level of circulation. Compression may not be tolerated and/or may have to be modified if arterial circulation is compromised. Arterial obstruction in the presence of venous insufficiency may not be readily recognized.

The ABI is the result of a vascular diagnostic test comparing the systolic blood pressure between the ankle and brachial pulses. An index result of 1.0 indicates an adequate arterial blood supply; an index less than 1.0 indicates insufficient blood flow to the distal regions for healing to occur (see Box 12-15, which includes acceptable values for compression therapy).

Assessing ABI is also warranted if wounds associated with CVI do not demonstrate healing within 2 weeks of beginning wound care. ABIs can be higher

than 1.0 in individuals with diabetes as the vessels do not compress due to arterial calcification.⁸

An assessment of the legs should be performed frequently to observe for insufficiency (stasis) ulcers, skin changes (e.g., color, texture, temperature), impaired growth of nails, and discrepancy in size of extremities, including observations and measurements for edema.

In the home health setting, the client or family should be instructed to contact a member of the medical team if any edema or change in the condition of the extremity occurs. When a stasis ulcer of any size is detected, treatment is initiated. A wound care specialist (usually a physical therapist or a nurse) is a vital part of the health care team in the management of stasis ulcers. Information on specific wound care management is available elsewhere.^{155,181,321,322}

Whirlpool beyond an initial one or two treatments is contraindicated, because the increased blood volume and dependent position (underlying causes of wound) can make the edema worse. When pulsatile debridement devices are unavailable, limited hydrotherapy (maximal temperature 80° F [26.7° C]) may be indicated to remove loose debris, and antiseptics may be indicated to moisten dried exudate or to facilitate debridement.

The client should be advised to avoid prolonged standing and sitting; crossing the legs; sitting too high for feet to touch the floor or too deep, causing pressure against the popliteal space; and wearing tight clothing (including girdles, elastic waistbands, or too-tight jeans) or support hose or stockings that extend above the knee, which act as a tourniquet at the popliteal fossa. Elastic stockings are recommended, but they must be worn properly to avoid bunching behind the knee or uneven compression in the popliteal fossa.

Vasomotor Disorders

Vasomotor disorders of the blood vessels causing headaches and reflex sympathetic dystrophy (now classified as complex regional pain syndrome) are discussed in Chapters 37 and 39, respectively.

Raynaud's Disease and Raynaud's Phenomenon

Definition and Overview. Intermittent episodes of small artery or arteriole constriction of the extremities causing temporary pallor and cyanosis of the digits (fingers more often than toes) and changes in skin temperature are called **Raynaud's phenomenon**. These episodes occur in response to cold temperature or strong emotion, such as anxiety or excitement. When this condition is a primary vasospastic disorder it is called (idiopathic) **Raynaud's disease**. If the disorder is secondary to another disease or underlying cause, the term Raynaud's phenomenon is used.

Incidence and Etiologic Factors

RAYNAUD'S DISEASE. Eighty percent of persons with Raynaud's disease are women between the ages of 20 and 49 years. The exact etiology of Raynaud's disease remains unknown, but it appears to be caused by hypersensitivity

of digital arteries to cold, release of serotonin, and genetic susceptibility to vasospasm. Raynaud's disease accounts for 65% of all people affected by this condition. Raynaud's disease is usually experienced as more annoying than medically serious.

RAYNAUD'S PHENOMENON. Epidemiologists estimate that Raynaud's phenomenon is a problem for 10% to 20% of the general population; it affects women 20 times more frequently than men, usually between the ages of 15 and 40 years. Risk factors for Raynaud's phenomenon are different between men and women. The Framingham Offspring Study reports that age and smoking are associated with Raynaud's phenomenon in men only, whereas an association with marital status and alcohol use was observed in women only. These findings suggest that different mechanisms influence the expression of Raynaud's phenomenon in men and women.¹¹²

Raynaud's phenomenon as a condition secondary to another disease is often associated with Buerger's disease or connective tissue disorders (collagen vascular diseases), such as Sjogren's syndrome, scleroderma, polymyositis and dermatomyositis, mixed connective tissue disease, SLE, and rheumatoid arthritis (see Box 12-17). Raynaud's phenomenon can be a sign of occult (hidden) neoplasm, especially suspected when it presents unilaterally.

Raynaud's phenomenon may also occur with change in temperature, such as occurs when going from a warm outside environment to an air-conditioned room. In addition, Raynaud's phenomenon may be associated with occlusive arterial diseases and neurogenic lesions, such as thoracic outlet syndrome, or with the effects of long-term exposure to cold (occupational or frostbite), trauma, or use of vibrating equipment such as jackhammers. Injuries to the small vessels of the hands may produce Raynaud's phenomenon. The trauma can be a result of repetitive stress that comes from using crutches for extended periods, typing on a computer keyboard, or even playing the piano.

Several medications (e.g., (3-blockers, ergot alkaloids prescribed for migraine headaches, antineoplastics used in chemotherapy) have also been implicated. Because nicotine causes small blood vessels to constrict, smoking can trigger attacks in persons who are predisposed to this phenomenon.

Pathogenesis and Clinical Manifestations. Scientists theorize that Raynaud's phenomenon is associated with a disturbance in the control of vascular reflexes. Although the causes differ for Raynaud's disease and Raynaud's phenomenon, the clinical manifestations are the same, based on a pathogenesis of arterial vasospasm in the skin.

It begins with the release of chemical messengers, which cause blood vessels to constrict and remain constricted. The flow of oxygenated blood to these areas is reduced, and the skin becomes pale and cold. The blood in the constricted vessels, which has released its oxygen to the tissues surrounding the vessels, pools in the tissues, producing a bluish or purplish color.

In the case of fibromyalgia-associated Raynaud's phenomenon, symptoms may be the result of cold-induced spasms of the arteries caused by a problem in the auto-

nomic nervous system control of the blood supply to the extremities. Altered or reduced numbers of α_2 -adrenergic receptors on the platelets correlate with Raynaud's phenomenon in fibromyalgia-associated Raynaud's.³⁵ These receptors are involved in the functioning of the autonomic nervous system. This could explain why the cold-induced pain is significant but without skin color changes in this population.

In most cases, the skin color progresses from blue to white to red. First, ischemia from vasospastic attacks causes cyanosis, numbness, and the sensation of cold in the digits (thumbs usually remain unaffected). The affected tissues become numb or painful. For unknown reasons, the flow of chemical that triggered the process eventually stops. The vessels relax, and blood flow is restored. The skin becomes white (characterized by pallor) and then red (characterized by rubor) as the vaso-spasm subsides and the capillaries become engorged with oxygenated blood. Oxygen-rich blood returns to the area, and as it does so, the skin becomes warm and flushed. The person may experience throbbing, paresthesia, and slight swelling as this occurs.

Sensory changes, such as numbness, stiffness, diminished sensation, and aching pain, often accompany vasomotor manifestations. Initially, no abnormal findings are present between attacks, but over time, frequent, prolonged episodes of vasospasm causing ischemia interfere with cellular metabolism, causing the skin of the fingertips to thicken and the fingernails to become brittle.

In severe, chronic Raynaud's phenomenon, the underlying condition may have produced scars in the vessels, reducing the vessel diameter and therefore blood flow. When attacks occur, they are often more severe, resulting in prolonged loss of blood to fingers and toes, which can produce painful skin ulcers; rarely, gangrene may develop. Episodes of Raynaud's disease are often bilateral, progressing distally to proximally along the digits. Raynaud's phenomenon may be unilateral, involving only one or two fingers, but this clinical presentation warrants a physician's differential diagnosis, since it can be associated with cancer (Fig. 12-35).

MEDICAL MANAGEMENT

DIAGNOSIS AND PROGNOSIS. Diagnosis is usually made by clinical presentation and past medical history. Raynaud's disease is diagnosed by a history of symptoms for at least 2 years with no progression and no evidence of underlying cause. Raynaud's disease must be differentiated from the numerous possible disorders associated with Raynaud's phenomenon. Untreated and uncontrolled Raynaud's may damage or destroy the affected digits. Rarely, necrosis, ulceration, and gangrene result. Even with intervention, the person with Raynaud's disease or phenomenon may experience disability and loss of function.

PREVENTION AND TREATMENT. Treatment for *Raynaud's disease* is limited to prevention or alleviation of the vasospasm because no underlying cause or condition has been discovered, although pharmacologic agents for primary and secondary Raynaud's phenomenon are



Figure 12-35

Raynaud's disease or phenomenon. White color (pallor) from arterio-spasm and resulting deficit in blood supply may initially involve only one or two fingers, as shown here. Cold and numbness or pain may accompany the pallor or cyanosis stage. Subsequent episodes may involve the entire finger and may include all the fingers. Toes are affected in 40% of cases. [From Jarvis C: *Physical examination and health assessment*, ed 4, Philadelphia, 2004, Saunders.]

under investigation. Clients are encouraged to avoid stimuli that trigger attacks, such as cool or cold temperatures, changes in temperature, and emotional stress, and to eliminate use of nicotine, which has a constricting effect on blood vessels.

Physical or occupational therapy is often prescribed and should include client education about managing symptoms through protective skin care and cold protection (see Box 12-14), biofeedback, stress management and relaxation techniques, whirlpool or other gentle heat modalities, and exercise. Large movement arm circles in a windmill fashion can restore circulation in some people. The individual will have to experiment with the speed at which to move the arms; some people benefit from slow, gentle movement, whereas others find greater success with fast rotations.

Calcium channel blockers are the treatment of choice. Pharmacologic management may also include nonaddicting analgesics for pain. Therapists can be instrumental in teaching physiologic modulation starting with hand warming. A hand-held device to measure fingertip temperature combined with self-guided or audio-guided relaxation can be very effective and is available.¹⁵⁶

When conservative care fails to relieve symptoms and the condition progresses clinically, sympathetic blocks followed by intensive therapy may be helpful. Sympathectomy may be necessary for persons who only temporarily benefit from the sympathetic blocks.

Treatment for *Raynaud's phenomenon* consists of appropriate treatment for the underlying condition or removing the stimulus causing vasospasm. The clinical care described for Raynaud's disease may also be of benefit. In addition, the use of antioxidants as an effective treatment of Raynaud's phenomenon as well as the role of therapeutic angiogenesis (regeneration of vessels) remains under investigation.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-23**Vasomotor Disorders****PREFERRED PRACTICE PATTERN**

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

Raynaud's Disease and Phenomenon

Prevention of episodes of Raynaud's is important. The affected individual must be encouraged to keep warm, avoid air conditioning, and dress warmly in the winter (e.g., protect the extremities as well as the head, chest, and back to maintain overall body temperature).

Aquatic therapy is often helpful in diminishing symptoms, but again, the individual must be careful when moving from place to place with extreme temperature changes (e.g., from outside winter temperatures into a warm pool area and back outside). The use of antihypertensives for Raynaud's can result in postural hypotension; the physician should be notified of these findings to alter the dosage.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-24**Peripheral Vascular Disease****PREFERRED PRACTICE PATTERNS**

4C: Impaired Muscle Performance

4J: Impaired Motor Function, Muscle Performance, Range of Motion, Gait, Locomotion, and Balance Associated with Amputation

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

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

7A: Primary Prevention /Risk Reduction for Integumentary Disorders (prevent complications of bed rest; other Integumentary patterns may apply depending on progression of disease and clinical manifestations)

Even though Special Implications for the Therapist boxes for each individual disease making up PVD have been presented, a brief overview or summary of PVD as a whole seems warranted and a reminder that because of the prevalence of atherosclerotic disease in anyone with PVD, heart rate and blood pressure should be monitored during the evaluative process and during initial interventions. This is especially important in those with diabetes mellitus and anyone who has undergone an amputation, which implies severe disease.

Notably, people with PAD may exhibit precipitous rises in blood pressure during exercise owing to the atherosclerotic process present and the diminished vascular bed.³⁴⁵ Examination of the pedal pulses should be part of the physical examination for all clients older than 55 years, and measurement of the ABI is recom-

mended for those who have diminished or nonpalpable pedal pulses but who do not have diabetes.⁸

For the client with back pain, buttock pain, or leg pain of unknown or previously undiagnosed cause, screening for medical disease, including assessment of risk factors, past medical history, and special tests and measures (e.g., bicycle test, palpation of pulses), is essential.^{129,133}

PVD can be confusing, with the wide range of diseases affecting veins and arteries, the etiology of which is sometimes occlusive, sometimes inflammatory, and occasionally, as in the case of Buerger's disease, both occlusive and inflammatory. The basic point to keep in mind is how arterial disease differs (significantly) from venous disease in clinical presentation, pathogenesis, and management.

Focusing on the underlying etiologic factors is the key to choosing the most appropriate and effective intervention. For example, in the case of acute arterial disease, the tissues are not oxygenated, and ischemia can result in local trauma or burns; gangrene can develop quickly. The goal is to increase oxygen without increasing demand or need for oxygen. Claudication occurs when the activity causes increased oxygen demand in an already compromised area.

During the acute phase of arterial ischemia rehabilitation, intervention and movement are minimized, heat and massage are contraindicated, and the person is instructed in the use of positions that will increase blood flow to the tissues involved (e.g., head elevated with legs slightly lower than the heart).

Chronic arterial disease can be treated by the therapist by concentrating on improving collateral circulation and increasing vasodilation. The role of exercise in PAD (especially in reducing claudication) has been well documented⁴⁶ (see also the section on Arterial Occlusive Diseases in this chapter). There is a suggestion that supervised exercise may be more beneficial than nonsupervised exercise.¹⁹⁶

In venous disorders, the tissues are oxygenated but the blood is not moving, and stasis occurs. With venous occlusion, the skin is discolored rather than pale (ranging from angry red to deep blue-purple), edema is prominent, and pain is most marked at the site of occlusion, although extreme edema can render all the skin of the limb quite tender.

The goal of therapy is to create compressive pumping forces to move fluid volume and reduce edema. For this reason, heat or cold, compressive stockings, massage, and activity (e.g., ankle pumps, heel slides, quad sets, ambulation) are part of the treatment protocol.

Further guidelines for exercise in the management of PVD are outlined elsewhere.^{11,139} See also the previous section on Arteriosclerosis Obliterans (Peripheral Arterial Disease). Modifying cardiovascular risk factors, improving exercise duration and decreasing claudication, preventing joint contractures and muscle atrophy, preventing skin ulcerations, promoting healing of any pressure ulcers, and improving quality of life are part of the therapy plan of care. In the case of lower extrem-

Continued.

ity amputation, the use of unweighted ambulation to reduce the physiologic demands of walking during early rehabilitation has been reported.²³¹

For people with vascular ulcers, improving the arterial supply or venous return will lessen pain, increase mobility, and allow ulcers to heal. Whenever ulcers are present, understanding the type of ulcer and underlying etiology will point to the best intervention. The assessment of and therapeutic intervention for vascular wounds are beyond the scope of this text; the reader is referred to other more appropriate texts.^{181,321,322}

Vascular Neoplasms

Malignant vascular (i.e., involving the blood vessels) neoplasms are extremely rare and include angiosarcoma, hemangiopericytoma, and Kaposi's sarcoma. *Angiosarcomas* (hemangiosarcomas) can occur in either gender and at any age, most commonly appearing as small, painless, red nodules on the skin, soft tissue, breast, bone, liver, and spleen. Almost one half of all people with angiosarcoma die of the disease.

Hemangiopericytoma arises from the smooth muscle cells that are external to the walls of capillaries and arterioles. Most commonly located on the lower extremities and retroperitoneum (space between the peritoneum lining the walls of the abdominal and pelvic cavities and the posterior abdominal wall), these tumors are composed of spindle cells with a rich vascular network. Metastasis to the lungs, bone, liver, and lymph nodes occurs in 10% to 50% of cases, but the majority of hemangiopericytomas are removed surgically without having invaded or metastasized.

Kaposi's sarcoma in association with AIDS most likely occurs as a result of loss of immunity. One form of the tumor resembles a simple hemangioma with tightly packed clusters of capillaries, most often visible on the skin. Although Kaposi's sarcoma is malignant and may be widespread in the body, it is not usually a cause of death.

Arteriovenous Malformations

Arteriovenous malformations (AVMs) are congenital vascular malformations of the cerebral vasculature. AVMs are the result of localized maldevelopment of part of the primitive vascular plexus consisting of abnormal arteriovenous communications without intervening capillaries. There is a central tangled mass of fragile, abnormal blood vessels called the *nidus* that shunts blood from cerebral feeding arteries directly into cerebral veins. The loss of the normal capillary network between the high-pressure arterial system and the low-pressure venous system results in a faster flow and elevated pressure within the delicate vessels of the AVM. The lack of a gradient pressure system predisposes the lesion to rupture.

AVMs vary in size, ranging from massive lesions that are fed by multiple vessels to lesions too small to identify. Perfusion to adjacent brain tissue may be impaired because blood flow is diverted to the AVM, a phenome-

non referred to as *vascular stealing*. AVMs may occur in any blood vessel, but the most common sites include the brain, GI tract, and skin. Approximately 10% of cases present with aneurysms. Small AVMs are more likely to bleed than large ones, and once bleeding occurs, repeated episodes are likely.

Clinical presentation depends on the location of the malformation and may relate to hemorrhage from the malformation or an associated aneurysm or to cerebral ischemia caused by diversion or stasis of blood. Seizures, migrainelike headaches unresponsive to standard therapy, and progressive neurologic deficits may develop.

Diagnostic testing and planned intervention rely on cerebral angiography to show the AVM size, location, feeding vessels, nidus, and venous outflow vessels. Other tests may include MRI, x-rays, ultrasound, electroencephalogram (EEG), and arteriogram. Treatment options are individualized depending on the size and location of the lesion as well as any other surgical risks present.

In the last 15 years, endovascular embolization and stereotactic radiosurgery (delivery of extremely precise doses of radiation to destroy abnormal blood vessels) have increased survival outcomes, especially for lesions previously considered inoperable or in cases of high surgical risk factors. Prognosis is guarded, since there is a 2% to 4% chance of hemorrhage with the concomitant risk of permanent neurologic deficit or even death.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-25

Arteriovenous Malformations

Generally, the individual with a known AVM is advised to avoid activities and exercise that can increase intracranial or blood pressure (see Box 16-1). Weight training and contact sports are contraindicated, and some physicians advise against high-aerobic exercise, including running.

Postoperative complications can include hemorrhage, seizures, nausea, vomiting, or headache, and symptomatic perilesional edema can occur up to 1 year after the procedure. Neurologic deficits vary but are usually transient; radiation-induced brain injury is rare. The radiation's effect begins immediately, but complete obliteration of the lesion can take up to 3 years, during which time the affected individual must continue to maintain a normal blood pressure.

The Lymphatic Vessels

The lymphatic system (see also the section on The Lymphatic System in Chapter 13) is part of the circulatory system that collects excess tissue fluid and plasma that has leaked out of capillaries into the interstitial space and returns it to the bloodstream.

The lymphatic system consists of lymphatic vessels and lymph nodes and functions to remove impurities from the circulatory system and to produce cells of the immune system (lymphocytes) that are vital in fighting bacteria and viruses. The lymph nodes are also part of the

lymphoid system, the organs and tissues of the immune system. All the lymphoid organs link the hematologic and immune systems in that they are sites of residence, proliferation, differentiation, or function of lymphocytes and mononuclear phagocytes (mononuclear phagocyte system).

Disorders of the lymphatic system may result from inflammation of a lymphatic vessel (lymphangitis), inflammation of one or more lymph nodes (lymphadenitis), an increased amount of lymph (lymphedema), or enlargement of the lymph nodes (lymphadenopathy). There are also three forms of lymph vascular insufficiency that can occur.

The first, *dynamic insufficiency*, occurs when the lymphatic load exceeds the lymphatic transport capacity. In this situation, the anatomy of the lymphatic system and its function are normal but are overwhelmed. A second form of insufficiency of the lymph vascular system is caused by a reduction of the lymphatic transport capacity below the level of a normal lymphatic protein load. This reduction results in low lymph flow failure called *mechanical insufficiency*.

A third form of lymph vascular insufficiency occurs when the lymphatic system has a reduced transport capacity, leading to an overflowing of lymph. This form is called *safety valve insufficiency*. For a complete discussion of the lymphatic system, see Chapter 13.

OTHER CARDIAC CONSIDERATIONS

Despite the success of new immunosuppressive regimens and better results with transplantation, few people who are dying of heart failure will actually have the opportunity to receive a heart transplant. The mechanical technology may eventually allow selected persons to receive long-term (permanent) support as a substitute for cardiac transplantation. The recipient is often home in 6 weeks with follow-up home health care. In the future, studies may be done to determine the efficacy of removing the cardiac assistive device after prolonged heart rest provides cardiac recovery. Survival with the natural recovered heart may be possible in some people.

Researchers are actively pursuing tissue engineering to replace transplantation and mechanical devices (e.g., artificial heart, implanted cardiac assistive devices or other bridges to transplantation) to help keep people alive while they await heart transplant or as a replacement intervention for transplantation.

The Cardiac Client and Surgery

Persons with previously diagnosed cardiac disease undergoing general or orthopedic surgery are at risk for additional postoperative complications. Anesthesia and surgery are often associated with marked fluctuations of heart rate and blood pressure, changes in intravascular volume, myocardial ischemia or depression, arrhythmias, decreased oxygenation, and increased sympathetic nervous system activity. In addition, changes in medications, surgical trauma, wound healing, infection, hemorrhage, and pulmonary insufficiency may overwhelm the

diseased heart. All these factors place an additional stress on the cardiac client during the perioperative period.

Cardiac surgery via median sternotomy requires a longitudinal incision and disruption of the sternum. During the operative procedure, the bone is rewired with stainless steel wire and fixed with low separation strength and security of closure approximately 5% of normal (this increases to 90% of normal strength at 6 weeks for most people).²¹⁴

A single-lung transplantation requires a posterolateral thoracotomy, whereas a double-lung transplant requires bilateral anterior thoracotomies referred to as a "clam shell." In this latter procedure, the rib cage and sternum are lifted perpendicularly as the hood of a car would be lifted. The heart-lung procedure is still done by open sternotomy.

Complications following a sternotomy include mediastinitis, poor wound healing, chronic pain, posttraumatic stress disorder, and, more rarely, brachial plexus injury. Risk factors for these complications may include obesity, osteoporosis, diabetes or other comorbidities, large breasts in women (the weight of both breasts puts additional traction on sutures), and client noncompliance or poor compliance.

Development of a less invasive means of performing cardiac surgery may be possible with recent advances in technology, especially videoscopic visualization and the ability to provide myocardial protection. Surgeons are examining alternate techniques in hopes of reducing operative stress, postoperative pain, and postoperative recovery time.

New procedures are being developed that eliminate the use of a sternotomy, such as the minithoracotomy or "keyhole" thoracotomy via a small incision that allows surgeons to operate on a beating heart. These alternate surgical techniques involve passing instruments through small incisions in the skin and muscle and between the ribs. Surgeons can suture bypass vessels around blocked coronary arteries without shutting down the heart and rerouting the blood through a bypass machine.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-26

The Cardiac Client and Surgery

Noncardiac Surgery

Therapy for people with cardiovascular disease undergoing orthopedic surgery or neurosurgery is only altered by the need for more deliberate and careful monitoring of the person's response to activity and exercise.

Postoperative rehabilitation may take longer because of the underlying cardiac condition and any complications that may arise as a result of cardiovascular compromise. Careful observation for DVT must be ongoing during the first 1 to 3 weeks postoperatively. Anyone with polycythemia or thrombocytopenia is at increased risk for hemorrhage, necessitating additional special precautions (see Chapter 14).

Continued.

Physical therapy initiated in the intensive care unit focuses on restoring mobility, increasing strength, and improving balance and reflexes; heel slides, ankle pumps, and bedside standing are included in the early postoperative protocol. Airway clearance techniques (formerly, chest physical therapy, pulmonary physical therapy, pulmonary hygiene) and breathing exercises are essential to prevent atelectasis (particularly left lower lobe atelectasis), especially in the case of implantation of an artificial heart, because of the location of the device. Frequent, slow, rhythmic reaching, turning, bending, and stretching of the trunk and all extremities many times throughout the day help alleviate the surgical pain-tension cycle and facilitate pulmonary function.

Cardiac Surgery

Progressive ambulation can be initiated as soon as the client can transfer. In the case of open heart surgery, sternal precautions are standard postoperative orders (see Box 12-3; see also Special Implications for the Therapist: Lung Transplantation in Chapter 21); preventing separation of the sternum may require hand-held assistance in place of assistive devices (e.g., walker, quad cane) initially. It is important to know whether the chest has been closed; the skin may be closed, but the chest may not be.

Upper extremity precautions are determined by the physician according to the surgery that was performed and the status of the incision. When the chest is closed, shoulder flexion and abduction can proceed until the point of movement at the chest wall or rib cage. This rotation can cause a torque, and further motion must be limited at that point.

When the client can ambulate 1000 feet, the treadmill (1 mile/hr) or exercise cycle (0.5 RPE) can be used (see Table 12-13), usually around the fourth postoperative day if there are no complications. Whether to use the treadmill or bicycle is generally an individual decision made by the client based on personal preference; presence of orthopedic problems must be taken into consideration. Resistive elastic or small weights and aerobic training are introduced between 4 and 6 weeks postoperatively. Pushing or pulling activities and lifting more than 10 lb are contraindicated in the first 4 to 6 weeks.

Chest (and in women, breast) discomfort, shortness of breath, upper quadrant myalgia (chest, arms, neck, upper back), palpitations, low activity tolerance, mood swings, and localized swelling in the case of grafts taken from the leg are all commonly reported in the early days and weeks after cardiac surgery. These clinical manifestations are minimized but not completely eliminated with the less invasive keyhole (minithoracotomy) surgery performed in some facilities.

Exercise

The use of lower extremity-derived aerobic exercise to improve hemodynamics, normalize heart rate, improve oxygen uptake and delivery, and decrease diastolic blood pressure has been well documented and discussed earlier in this chapter. Many of these individu-

als have not exercised in years and remain deconditioned or fearful of exercise.

The therapist must firmly encourage active participation in a program of physical activity and exercise for anyone who has given up and chosen to remain sedentary. Exercise tolerance must be monitored closely during the early weeks after surgery. The therapist is encouraged to use perceived exertion scales, such as the dyspnea index or Borg Scale (see Table 12-13), monitor changes in diastolic pressure, and rely on measurements of oxygen uptake to set exercise limits.

Psychosocial Considerations

Psychologic and emotional recovery from cardiac surgery is not always addressed or discussed. Recent research has documented that cardiac surgery is often accompanied by significant cognitive decline, especially memory loss (verbal and visual) and decline in task planning ability (visuoconstruction) and psychomotor speed.

Additional research is needed to determine if the observed cognitive decline is related to the surgery itself (e.g., effects of anesthesia, hypoperfusion associated with use of the heart-lung bypass machine, disruption of atherosclerotic plaque-forming emboli), normal aging in a population with cardiovascular risk factors, or a combination of these and other factors.^{299,301}

Depression is commonly reported after CABG and after cardiac surgery in general. The majority of people who are depressed after cardiac surgery were depressed before surgery. There does not appear to be any correlation between depressed mood and cognitive decline after cardiac surgery, which suggests that depression alone cannot account for the cognitive decline.

Since cardiac surgery is increasingly performed in older adults with more comorbidities, identifying people at risk for adverse neurocognitive outcomes will be helpful in protecting them by modification of the surgical procedure or by more effective medical therapy.^{216,299,301}

Cardiogenic Shock

Shock is acute, severe circulatory failure associated with a variety of precipitating conditions. Regardless of the cause, shock is associated with marked reduction of blood flow to vital organs, eventually leading to cellular damage and death. See Table 14-1 for categories and causes of shock.

The therapist may see a client in one of three stages of shock. *Stage 1*, compensated hypotension, is characterized by reduced cardiac output that stimulates compensatory mechanisms that alter myocardial function and peripheral resistance. During this stage, the body tries to maintain circulation to vital organs such as the brain and the heart and clinical symptoms are minimal. Blood pressure may remain normotensive.

In *stage 2*, compensatory mechanisms for dealing with the low delivery of nutrients to the body are over-

whelmed, and tissue perfusion is decreased. Early signs of cerebral, renal, and myocardial insufficiency are present. Cardiogenic shock (inadequate cardiac function) may result from disorders of the heart muscle, valves, or electrical pacing system. Shock associated with MI or other serious cardiac disease carries a high mortality rate. The therapist is only likely to see this type of client in a CCU setting.

Stage 3 is characterized by severe ischemia with damage to tissues by toxins and antigen-antibody reactions. The kidneys, liver, and lungs are especially susceptible; ischemia of the GI tract allows invasion by bacteria with subsequent infection.

Clinical manifestations of shock may include (in early stages) tachycardia, increased respiratory rate, and distended neck veins. In early septic shock (vascular shock caused by infection), there is hyperdynamic change with increased circulation, so that the skin is warm and flushed and the pulse is bounding rather than weak.

In the second phase of shock (late septic shock) hypoperfusion (reduced blood flow) occurs with cold skin and weak pulses, hypotension (systolic blood pressure of 90 mm Hg or less), mottled extremities with weak or absent peripheral pulses, and collapsed neck veins. This phase is usually irreversible; the client is unresponsive, and cardiovascular collapse eventually occurs. The therapist should be aware that some healthy adults may have blood pressure levels this low without ill effects or with only minor symptoms of orthostatic hypotension when changing positions quickly.

Treatment is directed toward both the manifestations of shock and its cause.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-27

Cardiogenic Shock

PREFERRED PRACTICE PATTERN

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

The therapist in an acute care or home health setting may be working with a client who is demonstrating signs and symptoms of impending shock. Careful monitoring of vital signs and clinical observations will alert the therapist to the need for medical intervention (see early signs of shock listed in previous section). The client in question may demonstrate normal mental status or may become restless, agitated, and confused.

For the acute care therapist, people hospitalized with shock are critically ill and are usually unresponsive. Cardiopulmonary and musculoskeletal function as well as prevention of further complications will be the focus of the therapist. Treatment for the immobile person in shock, which is directed toward positioning, skin care, and pulmonary function, must be short in duration but effective, to avoid fatiguing the person.

The Cardiac Client and Pregnancy

Normal physiologic changes during pregnancy can exacerbate symptoms of underlying cardiac disease, even in previously asymptomatic individuals. The most common cardiovascular complications of pregnancy are peripartum cardiomyopathy, aortic dissection, and pregnancy-related hypertension.

Peripartum cardiomyopathy or cardiomyopathy of pregnancy is discussed briefly earlier in the chapter (see the section on Cardiomyopathy). Pregnancy predisposes to aortic dissection, possibly because of the accompanying connective tissue changes. Dissection usually occurs near term or shortly postpartum in the arteries (including coronary arteries) or the aorta, and special implications are the same as for aneurysm.

The Heart in Collagen Vascular Diseases

Collagen vascular diseases (now more commonly referred to as diffuse connective tissue disease) (Box 12-17) often involve the heart, although cardiac symptoms are usually less prominent than other manifestations of the disease.

Lupus Carditis

SLE is a multisystem clinical illness (see Chapter 7) characterized by an inflammatory process that can target all parts of the heart, including the coronary arteries, pericardium, myocardium, endocardium, conducting system, and valves. Lupus cardiac involvement may include pericarditis, myocarditis, endocarditis, or a combination of the three. Cardiac disease can occur as a direct result of the autoimmune process responsible for SLE or secondary to hypertension, renal failure, hypercholesterolemia (excess serum cholesterol), drug therapy for SLE, and, more rarely, infection (infective carditis).

Pericarditis is the most frequent cardiac lesion associated with SLE, presenting with the characteristic substernal chest pain that varies with posture, becoming worse in recumbency and improving with sitting or bending forward. In some people, pericarditis may be the first manifestation of SLE.

Myocarditis (see also the section on Myocardial Disease) is a serious complication reported to occur in less than

Box 12-17

COLLAGEN VASCULAR DISEASES

- Ankylosing spondylitis
- Dermatomyositis
- Localized (cutaneous) scleroderma
- Mixed connective tissue disease
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Polymyositis
- Rheumatoid arthritis (RA)
- Sjögren's syndrome
- Systemic lupus erythematosus (SLE)
- Systemic sclerosis (scleroderma)
- Temporal arteritis

10% of people with SLE. The simultaneous involvement of cardiac and skeletal muscle may occur more commonly than previously suspected. More sensitive diagnostic techniques now make early detection of occult myocarditis possible. Myocarditis in association with SLE occurs most often as left ventricular dysfunction and conduction abnormalities with varying degrees of heart block.

Lupus *endocarditis* occurs in up to 30% of persons affected by SLE. Major lesions associated with lupus endocarditis include the formation of multiple noninfectious wartlike elevations (verrucae) around or on the surface of the cardiac valves, most commonly the mitral and tricuspid valves. Other types of valvular disease associated with SLE include mitral and aortic regurgitation or stenosis.

Rheumatoid Arthritis

On rare occasions, the heart is involved as a part of rheumatoid arthritis, a chronic, systemic, inflammatory disorder that can affect various organs but predominantly involves synovial tissues of joints (see Chapter 27). When the heart is affected, rheumatoid granulomatous inflammation with fibrinoid necrosis may occur in the pericardium, myocardium, or valves. Involvement of the heart in rheumatoid arthritis does not compromise cardiac function.

Ankylosing Spondylitis

Ankylosing spondylitis is a chronic, progressive inflammatory disorder affecting fibrous tissue primarily in the sacroiliac joints, spine, and large peripheral joints (see Chapter 27). A characteristic aortic valve lesion develops in as many as 10% of persons with longstanding ankylosing spondylitis. The aortic valve ring is dilated, and the valve cusps are scarred and shortened. The functional consequence is aortic regurgitation (see the section on Aortic Regurgitation [Insufficiency]).

Scleroderma

Scleroderma or systemic sclerosis is a rheumatic disease of the connective tissue characterized by hardening of the connective tissue (see Chapter 10). Involvement of the heart in persons with scleroderma is second only to renal disease as a cause of death in scleroderma. The myocardium exhibits intimal sclerosis (hardening) of small arteries, which leads to small infarctions and patchy fibrosis. As a result, CHF and arrhythmia are common. Cor pulmonale may occur secondary to interstitial fibrosis of the lungs, and hypertensive heart disease may occur as a result of renal involvement.

Polyarteritis Nodosa

Polyarteritis refers to a condition of multiple sites of inflammatory and destructive lesions in the arterial system; the lesions consist of small masses of tissue in the form of nodes or projections (nodosum) (see previous discussion in this chapter). The heart is involved in up to 75% of cases of polyarteritis nodosa. The necrotizing lesions of branches of the coronary arteries result in MI, arrhythmias, or heart block. Cardiac hypertrophy and failure secondary to renal vascular hypertension occur.

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-28

Collagen Vascular Diseases

Treatment of the collagen vascular diseases described must take into consideration the possibility of cardiac involvement. The physician has usually diagnosed concomitant cardiac disease, but complete health care records are not available to the therapist. If the therapist identifies signs or symptoms of cardiac origin, the client may be able to confirm previous diagnosis of the condition. In such cases, careful monitoring may be all that is required. However, the alert therapist may be the first health care provider to identify signs or symptoms of underlying dysfunction during onset, necessitating medical referral. (See each collagen vascular disease for discussion of individual implications.)

Cardiac Complications of Cancer and Cancer Treatment²²⁷

Many treatments for cancers are also known to be cardio-toxic. People with cancer experience all the usual cardiac problems that occur in the general population in addition to complications of cancer and its therapy. Tumor masses can cause compression of the heart and great vessels resulting in pericardial effusions and tamponade. Certain tumors can cause arrhythmias and may secrete mediators that are directly toxic to the heart. Pericardial effusions and tamponade can follow surgery, radiation, or chemotherapy.

Cardiac toxicity may occur following chest irradiation, especially when combined with the administration of many chemotherapeutic agents. Chest radiation for any type of cancer (e.g., Hodgkin's disease, non-Hodgkin's lymphoma, esophageal cancer, lung cancer, breast cancer) exposes the heart (and lungs) to varying degrees and doses of radiation. Previous mediastinal radiation and increasing cumulative doses of chemotherapy or irradiation are known risk factors for the development of cardiotoxicity.

Radiation exposure can cause considerable scarring within the subendocardial adipose tissue, endocardial thickening, and interstitial fibrosis.²⁸⁴ Collectively the latter three defects would make the heart less capable of expanding during systole. Pericardial effusion is the most common manifestation of radiation heart disease, but coronary arteries are known to become fibrotic and undergo luminal narrowing, resulting in hypertension, angina, and MI.

Doxorubicin (Adriamycin) was identified as being cardiotoxic during the early drug trials in the 1970s, but it took 5 to 6 years of actual use for the full extent of cardiac damage to become obvious. Today, cumulative doses of doxorubicin are limited to approximately 550 mg/m² because the drug can cause fatal CHF in doses above this amount. Often these effects are not seen until years or decades after treatment with the drug has been com-

pleted. Many other chemotherapeutic agents are cardiotoxic but not to the extent of doxorubicin, and these effects tend to be more acute than chronic.³⁶⁶

Chemotherapy agents may prompt acute and chronic heart failure (e.g., anthracycline antibiotics, mitoxantrone, doxorubicin combined with paclitaxel used in the treatment of breast cancer)²⁶² or coronary spasm leading to angina, MI, arrhythmias, or sudden death (e.g., 5-fluorouracil). Anthracycline effects on the heart reduce exercise tolerance. Endocarditis also occurs in cancer clients with vascular access devices and immune compromise.

Recombinant technology has resulted in the development of biologic response modifiers, including the interferons, interleukins, and tumor necrosis factor, which also have some adverse cardiovascular effects. Hypotension and tachycardia are the most common problems, although there have been some reports of myocardial ischemia and infarction. These adverse effects appear to be caused by significant alterations in fluid balance rather than any dysrhythmic or cardiotoxic properties of the drugs. Fortunately, many of the cardiac complications associated with chemotherapeutic agents and biologic response modifiers are transient and reversible.¹⁵⁰

The most common manifestations of cardiotoxicity are cardiac arrhythmias or acute or chronic pericarditis. Other cardiac problems that may develop include blood pressure changes, thrombosis, ECG changes, myocardial fibrosis with a resultant restrictive cardiomyopathy, conduction disturbances, CHF, accelerated and radiation-induced CAD, and valvular dysfunction. These may occur during or shortly after treatment or within days or weeks after treatment; or they may not be apparent until months and sometimes years after completion of chemotherapy.²⁵¹

Although only a small percentage of persons develop serious problems or obvious symptoms of cardiotoxicity, many people have functional limitations that are not clinically apparent because they are physically inactive. A number of risk factors may predispose someone to cardiotoxicity, including total daily dose, increasing cumulative dose, schedule of administration, concurrent administration of cardiotoxic agents, prior chemotherapy, mediastinal radiation, age (younger than 18 years or older than 70 years), female gender, history of preexisting cardiovascular disorders or other comorbidities such as diabetes, and presence of electrolyte imbalances (e.g., hypokalemia, hypomagnesemia).²⁵¹

SPECIAL IMPLICATIONS FOR THE THERAPIST 12-29

Cardiac Complications of Cancer Treatment

Any client referred to therapy who has completed oncologic treatment should be assessed for potential cardiac (and pulmonary) dysfunction, including questions about previous and current activity levels, evaluation of exercise tolerance or endurance, monitoring of heart rate and rhythm, blood pressure, and respiratory responses.

Any symptoms of exercise intolerance (shortness of breath, light-headedness or dizziness, fatigue, pallor, palpitations, chest pain or discomfort) must be noted. (See also Special Implications for the Therapist: Cardiomyopathy and Special Implications for the Therapist: Pericarditis.)

Clients may be asymptomatic, with the only manifestation being ECG changes. Ideally, the oncology and cardiac team will recommend continuous cardiac monitoring with baseline and regular ECG and echocardiographic studies and measurement of serum electrolytes and cardiac enzymes for those individuals with risk factors or a history of cardiotoxicity.

Specific exercise guidelines have also been outlined for the inclusion of gradual endurance training as a part of the treatment plan for anyone with cardiotoxicity secondary to oncologic treatment.³⁴⁵ (See also the sections on Radiation Injuries in Chapter 5 and Cancer and Exercise in Chapter 9.)

Cardiotoxicity can be prevented by screening and modifying risk factors, aggressively monitoring for signs and symptoms as chemotherapy is administered, and continuing follow-up after completion of a course or the entire treatment. Cardioprotective agents are being developed with approval by the FDA, such as dexrazoxane for anthracycline chemotherapy.²⁵¹

References

To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this textbook. The reader can view the reference source and access it on-line whenever possible. There are a total of 369 cited references and other general references for this chapter.

CHAPTER 13

The Lymphatic System

BONNIE B. LASINSKI

The lymphatic system develops embryologically from the venous system. Two major theories exist of the embryologic origin of the lymphatic system: the *centrifugal*, or venous budding, theory and the *centripetal* theory. The centrifugal theory states that the lymphatic endothelium develops from the venous endothelium; the centripetal theory states that both systems (venous and lymphatic) develop from undifferentiated (stem type) mesenchymal cells. Advances in lymphangiogenesis research may clarify which theory is correct; this information will have a great impact on genetic research and the eventual molecular treatment of lymphangiodyplasias.⁹²

The interstitial fluid that remains after the extracellular fluid is resorbed via the veins is taken up by the initial lymphatic vessels, into larger collecting vessels, into lymphatic trunks, and back into the right side of the heart via the lymphatic ducts that empty into the subclavian veins in the neck. This is a regional system that moves fluid from the periphery to the central circulation. It is designed to help maintain fluid balance in the tissues, fight infection, and assist in the removal of cellular debris and waste products from the extracellular spaces. In many ways, it functions like the "sanitation" system of a major city. It is largely ignored and goes unnoticed until it is disrupted and the "garbage" (in the form of lymphedema) piles up. This drainage system is separate from the general circulatory system but is the conduit for returning tissue fluids to the circulatory system.^{42,87}

The cardiovascular system is a one-cycle system of vessels with a pump (the heart) to move the fluid (blood) through arteries, capillaries, and veins and then back to the heart via the veins. Fluid that leaves the arterial side of the capillary bed in a process called *ultrafiltration* nourishes the tissues and cells.

Of the fluid volume that perfuses the tissues, 90% reenters the circulation via the venous capillary network (*reabsorption*) because of differences in concentration of fluid and protein in the tissues and in the venous end of the capillary network. The remaining 10% of extracellular tissue fluid and plasma proteins in the interstitial spaces must be returned to the heart via the lymphatic system.

Most protein molecule's diameters are too large to pass through openings in the endothelium of the venous capillaries. A small amount of protein, if broken down into smaller molecules by macrophages, can pass through the open junctions in the venous endothelium. However, the

majority of extracellular protein must be transported via the lymphatic system. Although 10% of the total fluid volume seems small, it can amount to 2 L/day.

The lymphatic system is a pressure-driven system based on the principles of osmotic diuresis. If the normal lymphatic transport mechanisms are disrupted (e.g., by scar tissue or reduced muscle pumping), significant accumulations of water and protein can remain in the tissue spaces, resulting in latent, acute, or chronic lymphedema. This protein is a result of cellular metabolism and is not related to protein ingested from food.

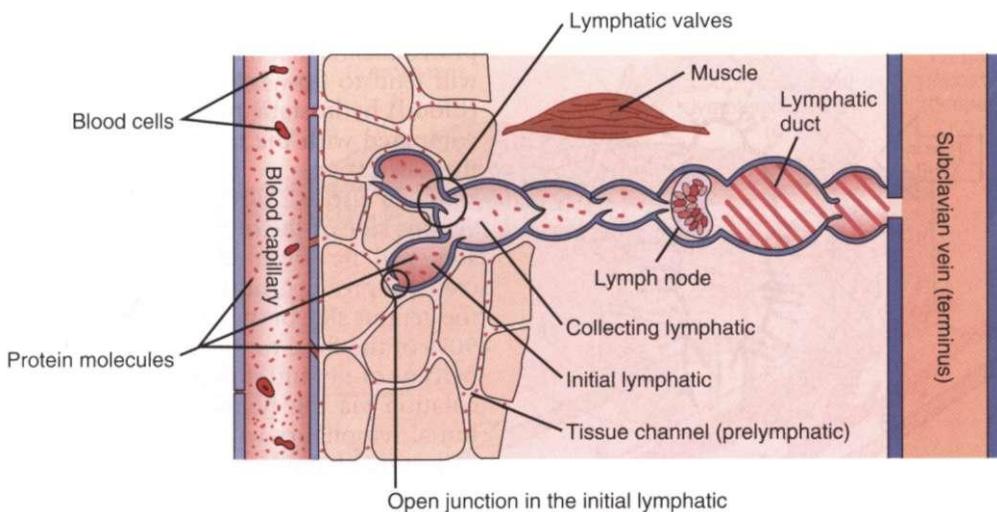
The dynamics of fluid exchange in the tissues are controlled by the microcirculation unit consisting of the arterial and venous capillaries, the tissue channels, the proteolytic cells (macrophages) in the tissues, and the initial lymphatics (see the description of initial lymphatics in the next section).

ANATOMY AND PHYSIOLOGY

The lymphatic system is composed of superficial and deep lymph vessels and nodes. Other lymphatic organs and tissues include the thymus, bone marrow, spleen, tonsils, and Peyer's patches of the small intestine. These perform important immune functions discussed in Chapter 7. Superficial vessels rely on an interaction of oncotic and hydrostatic pressures, muscle contraction, arterial pulsation, and gentle movement of the skin to move lymph fluid, whereas the deeper vessels, which generally parallel the venous system, contain smooth muscle and valves and help prevent backflow.¹⁹

The lymphatic vessel network is an intricate one-way vessel system that serves to drain the 10% excess tissue fluid volume and plasma proteins that remain in the interstitium after normal capillary perfusion/filtration has taken place and return it to the central circulation via the large veins in the neck. All lymph fluid eventually passes through lymph nodes before emptying into the right lymphatic duct and the thoracic duct. The fluid is then returned to the bloodstream via the left and right subclavian veins.^{42,87}

The anatomy of the lymphatic vessel system can be compared in some ways to the vein system on the leaves and stems of trees. The smallest vessels, or veins, are at

**Figure 13-1**

Anatomy of the lymphatic vessel system (schematic). This diagram shows the passage of protein (dots) in normal tissue from the blood capillary, through the tissue channels, into and through the lymphatic system, back to the venous system, and eventually emptying into the subclavian vein. Note that the protein molecules are not on the venous side of the diagram because for the most part, these molecules are too large to pass through the openings in the venous endothelium. Also note that this is a schematic diagram and not drawn to scale; the lymphatic duct depicted emptying into the venous system (subclavian vein) is much deeper (under the fascia) than this two-dimensional illustration can portray. Various malfunctions are illustrated in Fig. 13-15. (From Casley-Smith JR, Casley-Smith JR: *Modern treatment for lymphoedema*, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia.)

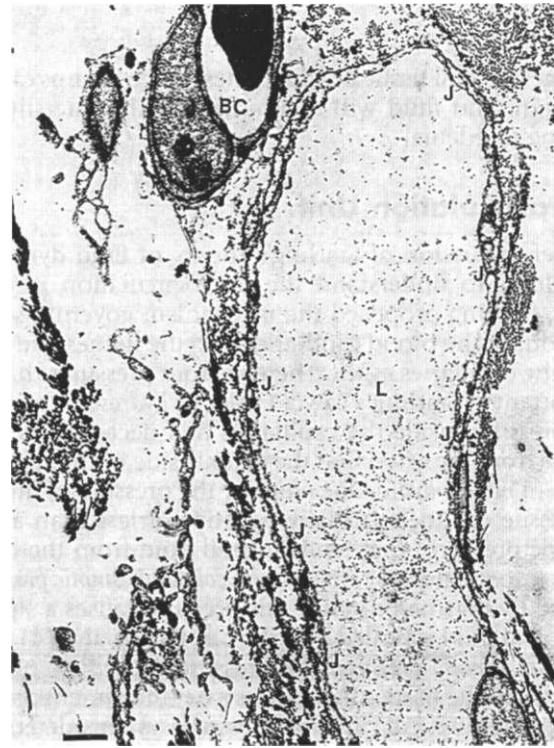
the periphery of the tissues (leaves), and the diameter of these vessels gradually increases in the stem of the leaf as the system progresses into larger and larger vessels (corresponding to deeper tissues) and continues to progress to larger stems and branches of the tree until the trunk is reached.

The smallest of lymphatic vessels (diameter 20 to 40 μm), called *lymphatic capillaries* or *precollectors*, begin as blind-ended sacs of endothelium just under the epidermis.^{19,32,42} These are referred to as the *initial lymphatics* and are in close proximity with the venous and arterial capillaries (Fig. 13-1). Terminology has changed over the years; the reader should be aware that initial lymphatic and lymphatic capillary refer to the same structure.

The vessel walls of these initial lymphatics are one cell thick, formed by overlapping endothelial cells with many loose junctions between cells opening and closing (Fig. 13-2). This action allows for movement of water and proteins into the vessel and prevents the escape of protein into the interstitium during the compression of the initial lymphatics.

These cells are also in direct contact with the microfilaments of the surrounding connective tissues. They are connected to the tissue matrix by anchoring filaments that act as "guide wires" to pull the cell junctions open when the tissue pressure rises as a result of increased extracellular fluid volume¹⁹ (Fig. 13-3). These vessels are arranged in a meshlike plexus; for every square millimeter of tissue, 7 mm of lymphatics are available to drain it.¹⁸

The initial lymphatics function as force-pumps powered by variations in total tissue pressure caused by movement, muscular contraction, respiration, and variations in external pressure caused by massage, gravity, change in position, and other similar factors. Without

**Figure 13-2**

An initial lymphatic (L) in a quiescent (at rest or inactive) tissue. Many closed (narrow or tight) junctions (J) are evident. A blood capillary (BC) is shown for comparison of size, endothelial opacity, and other characteristics. The bar at the lower left (1 μm) is provided to give the viewer size perspective. (From Casley-Smith JR, Casley-Smith JR: *Modern treatment for lymphoedema*, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia. Modified from Casley-Smith JR: *Br J Exp Path* 46:35-49, 1965.)

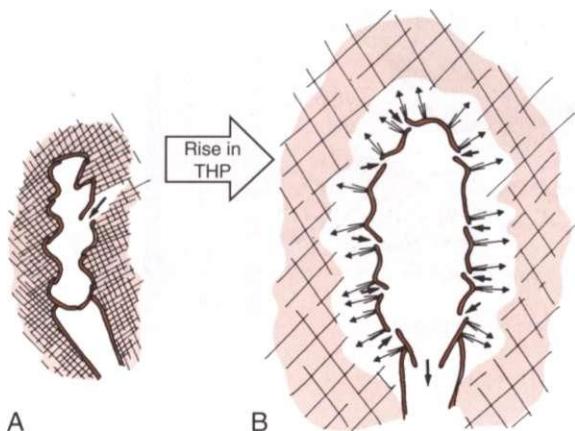


Figure 13-3

Effects of elevated tissue hydrostatic pressure (THP) on initial lymphatic functioning. **A**, Normal lymphatic vessel at a fairly low tissue hydrostatic pressure and normal lymphatic drainage. **B**, Tissue response to a tremendous increase in tissue hydrostatic pressure (represented by the large arrow). The swelling in the interstitial tissues pulls on the anchoring filaments, pulling and holding open the initial lymphatic endothelial junctions (thin arrows pointing outward), allowing fluid to pour into the initial lymphatic in an attempt to reduce edema. In this way, in place of a few widely open junctions, there are many slightly open ones, through which fluid is forced (thicker arrows directed inward) down a hydrostatic pressure gradient. (From Casley-Smith JR, Casley-Smith JR: *Modern treatment for lymphoedema*, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia.)

changes in total tissue pressure, these force-pumps cannot function, and fluid will accumulate in the interstitium, leading to edema.

Microcirculation Unit

A brief discussion of Starling's theory of fluid dynamics is helpful to understand the microcirculation unit. In 1897, Starling proposed the mechanism governing fluid flow out of the blood capillaries into the tissues and back into the capillaries again. There are four pressures that are important in Starling's Law: (1) *plasma hydrostatic pressure*, the pressure inside the capillaries that decreases as fluid passes from the arterial to the venous side of the capillary loop; (2) *tissue hydrostatic pressure*, the pressure of fluid in the tissue channels (usually negative or less than atmospheric pressure) that tends to pull fluid from the capillaries into the tissues*; (3) *plasma colloidal osmotic pressure*, caused by plasma proteins, this pressure causes a siphon effect and fluid is pulled into the capillaries; and (4) *tissue colloidal osmotic pressure*, the pressure caused by plasma proteins in the tissues that causes a net movement of fluid into the tissues. All of these pressure systems determine how much fluid moves and where it moves within the body.

The laws of basic fluid dynamics dictate that fluid flows from an area of high pressure to an area of lower

pressure until equilibrium is reached. Starling's law simplified means that fluid at the arterial end of the capillary will tend to flow into the tissue spaces because plasma (blood) hydrostatic pressure is higher at the arterial end compared with the tissue hydrostatic pressure (THP) of the tissues.

If all else is "normal," when the fluid reaches the venous side of the capillary, the plasma hydrostatic pressure will be lower than the plasma colloid (protein) osmotic pressure, and the fluid will be forced back into the venous side of the capillary, which accounts for about 90% of the fluid on the venous end of the capillary. Ten percent of net-ultrafiltrate must return to the central circulation via the lymphatics. If all is normal there, the initial lymphatics will take up that fluid, move it to the collecting lymphatics and larger lymphatic trunks, through regional lymph nodes, and eventually into the right lymphatic duct or the thoracic duct, and back into the vena cava (see Fig. 13-8).

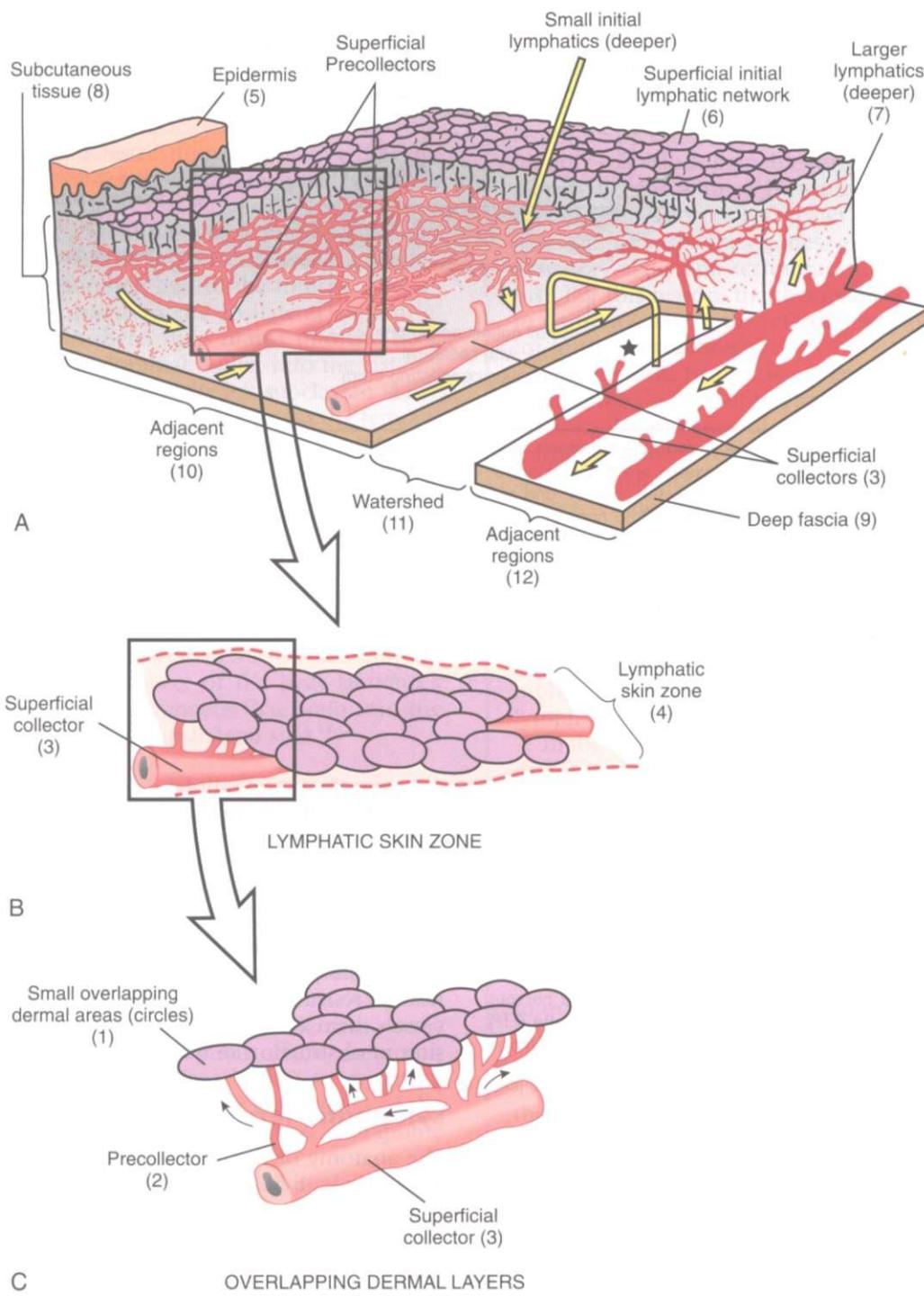
In the presence of lymphatic dysfunction, some part or all of the 10% of fluid volume and the proteins will remain trapped in the tissue spaces and cause lymphedema. With lymphedema, the challenge for the therapist is to effectively move that fluid back into functioning lymphatics and then into the central circulation. This model is a very gross simplification but can be helpful in understanding the basics of fluid dynamics in the capillary loop. Many other factors affect the tissues in addition to those mentioned previously.^{19,90}

Deeper in the dermis are *precollectors* (Fig. 13-4), which flow into *collecting lymphatics* located in the subcutaneous tissue (Fig. 13-5). The true collecting vessels have valves to prevent backflow and some muscle tissue in their walls to further enhance their pumping action.^{19,90} Extrinsic muscle contraction or lymphatic drainage massage also increases this pumping action.

Collecting lymphatics do not form a plexus, but there can be connections between them. Their diameter gradually increases in size to form the *lymph trunks*, which lie near the deep fascia. Each segment of collecting lymphatic vessels between valves is called a *lymphangion* (Fig. 13-6). The muscle in the collecting lymphatic walls contracts rhythmically. Smooth muscle cells around the endothelial cell layer face the lumen of the vessel. These are innervated by the autonomic nervous system and contract on the average of 5 to 10 times per minute.⁹³ This "lymphangiomotoricity" combines with the contraction of the lymphangion itself, which is triggered by distention of the vessel wall. The greater the stretch, the greater the force of the contraction. If many lymphangions contract at once and outflow is obstructed (e.g., by scarred or radiated lymph nodal areas), pressure inside the vessel can reach as high as 100 mm Hg.

High intravascular pressure fatigues the muscle wall, leading to ineffective smooth muscle contraction and ultimately to vessel failure. The walls dilate, preventing closure of valve flaps, and a backflow of lymph distal to the site of obstruction occurs, causing lymphedema. This is one plausible explanation for the fact that many individuals with a "limb at risk" develop lymphedema months or even years after their original surgery. For a time, the remaining lymphatics function marginally without

*However, in edema, it can become positive and tend to keep fluid out of the tissues. This is the one "safety factor" that controls lymphedema (i.e., the fibrous tissue actually helps to increase tissue hydrostatic pressure [THP] and control the size of the limb to some extent).

**Figure 13-4**

Overview of the lymphatic drainage system. **A**, Overview of the lymphatic drainage paths from a skin region. The epidermis (5) is superficial to a superficial initial lymphatic network (6), which sends blindly ending vessels into the dermis and which is linked to the deep dermal plexus of larger initial lymphatics (7), in the subcutaneous tissue (8), by many connections. The superficial collecting lymphatics (3), which discharge into the larger ones (not shown), lie next to the deep fascia (9). A watershed (11) lies between two adjacent regions (10 and 12), which drain in opposite directions (medium arrows). One of these is obstructed (red vessels). The deep and superficial initial lymphatic plexuses overlap across this watershed. These groups of cross-connections provide collateral drainage and are enlarged by manual massage. The large, U-shaped arrow (*) shows this path. **B**, Lymphatic skin zone (4) that extends along the length of a superficial collector (3). Certain areas of the skin drain into a specific superficial collector, which accounts for the clinical observation of lymphedema in portions of an extremity (e.g., pockets of extra swelling or asymmetric edema). When a specific superficial collector is blocked (or if the deep collector into which it drains is blocked), the result is edema at that site. **C**, Shows (1) small overlapping dermal areas (circles), which drain into networks of initial lymphatics (not shown), which drain into small collecting lymphatics called precollectors (2) and then to larger superficial collectors (3). (From Casley-Smith JR, Casley-Smith JR: Modern treatment for lymphoedema, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia. Modified from Földi M, Kubik S: Lehrbuch der lymphologie für mediziner und physiotherapeuten mit anhang: praktische linweise für die physiotherapie, Stuttgart, Germany 1989, Gustav Fischer Verlag.)

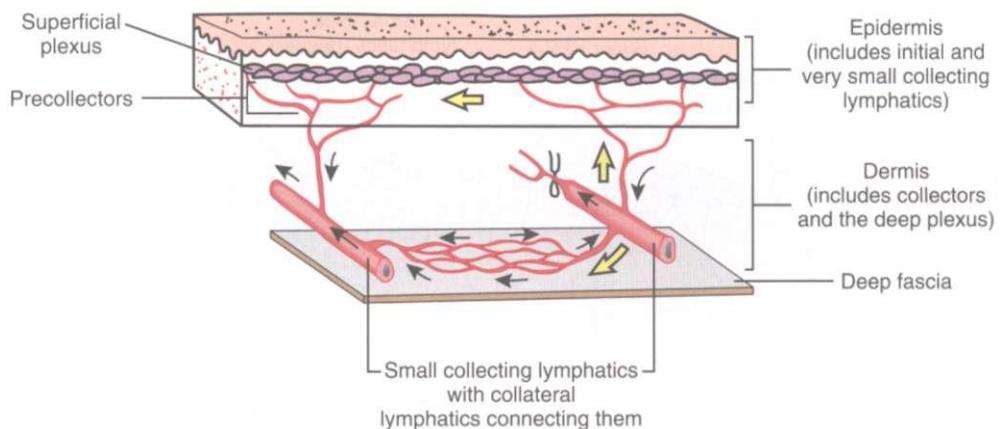


Figure 13-5

Collecting lymphatics. Lymphatics traverse through the epidermis, dermis, and deep fascia via lymphatics that increase in size as they go deeper into the tissues. Two layers of lymphatic plexuses are in the skin: the epidermis and dermis (layer just below the epidermis, formerly called the corium), which contains blood and lymphatic vessels, nerves, and nerve endings, glands, and hair follicles. Lymphatic vessels in the dermal layer can divert fluid from a blocked area and drain it into normally functioning area(s). In this illustration, one of the two larger collectors (right) is blocked; note the watershed between the blocked and the open collecting lymphatic. The lymph that would normally be transported along this blocked collector instead passes up into the superficial plexus and down into the deeper plexus formed by collaterals in the watershed area located just above the deep fascia. In these, the lymph passes to the nonblocked collector (left) and then drains into larger lymph vessels (not shown). When edema exists, the valve flaps in the collaterals are dilated and do not meet, thereby allowing lymph to move in either direction across these vessels (i.e., across the watershed). (From Casley-Smith JR, Casley-Smith JR: *Modern treatment for lymphoedema*, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia. Modified from Földi M, Kubik S: *Lehrbuch der lymphologie für mediziner und physiotherapeuten mit anhang: praktische linweise für die physiotherapie*, Stuttgart, Germany 1989, Gustav Fischer Verlag.)

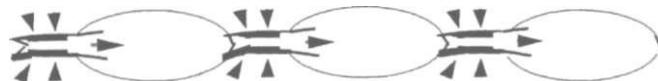


Figure 13-6

The lymphangion. Many lymphangions may contract at once, but sometimes only one lymphangion is triggered. The pressure exerted by each lymphangion is usually a few mm Hg but can be over 100 mm Hg if outflow is obstructed and many units are contracting at once. Contraction is triggered by distention (i.e., greater filling creates greater force) but can be modified by humoral (including medications) and nervous factors. Pumping is greatly aided by varying total tissue pressure (TPP) (e.g., from adjacent muscles, respiration, or manual lymphatic drainage [MLD]), as previously mentioned in the text. (From Casley-Smith JR, Casley-Smith JR: *Modern treatment for lymphoedema*, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia.)

evidence of clinical lymphedema, but these units become overtaxed, eventually the walls fatigue, and latent lymphedema progresses to acute and then to chronic lymphedema.

The lymph trunks in the extremities join into the larger lymph vessels of the trunk, which join to form the *thoracic duct* and the *right lymphatic duct* that pump the lymph into the central circulation at the left and right subclavian veins in the root of the neck. The lymphatic vessels are embedded in fatty tissue and accompany the chains of lymph nodes along the blood vessels.³² This explains why injury to blood vessels in an area implies injury to lymphatic vessels in that area, too, regardless of whether it is "unexpected" or "controlled" trauma, as in surgery.

As lymph flows from the periphery to the root of the limbs to the center of the body, it passes through many *lymph nodes*, which act as filters to cleanse the lymph of

waste products and cellular debris. Vessels distal to nodes are called *afferent lymph vessels*. Vessels leaving lymph nodes for more proximal points are called *efferent lymph vessels*.

Lymph nodes also produce lymphocytes and macrophages, which are critical for immune function; destroy foreign bacteria, harmful viruses, and cancer cells; and filter waste products. Lymph nodes offer 100 times the normal resistance to flow of lymph within the lymphatic vessels themselves, which explains why they are often the sites of obstruction in lymphatic dysfunction.^{19,40,42,90}

Lymphatic Territories and Watersheds

The anatomy of the lymphatic system is a regional one. The body is divided into a series of lymph drainage territories called *lymphotomes*, which are bordered and separated by so-called watershed areas. The watershed areas are characterized by sparse collateral flow to adjacent lymphotomes,^{11,41} but connections exist between lymphotomes in the superficial and deep plexuses and via collateral lymphatics between deep collectors in adjacent lymphotomes located just above the deep fascia. Under normal conditions, the lymph drains in different directions on either side of these watersheds (Fig. 13-7).

The trunk can be divided into four quadrants: the left and right thoracic lymphotomes and the left and right abdominal lymphotomes. The left and right thoracic lymphotomes drain into the ipsilateral axilla, as do the left and right upper extremities. Some individuals possess an auxiliary drainage pathway from the lateral aspect of the upper arm called the *deltoid-pectoral* or *cephalic chain*. This pathway drains directly into the ipsilateral

subclavian nodal area, bypassing the axilla entirely. If present, an individual may be less likely to develop upper extremity lymphedema secondary to axillary disruption (surgical or by radiation), as this pathway may provide sufficient lymph transport capacity for the upper extremity. This pathway can be disrupted by supraclavicular radiation therapy sometimes used to treat recurrent cancer of the chest wall.⁵⁴

The left and right abdominal lymphotomes drain into the left and right superficial inguinal nodes, respectively. Each leg and corresponding half of the lumbar, gluteal, and genital region drains to the ipsilateral superficial inguinal nodes. From there, fluid drains into the deep inguinal, pelvic, and abdominal nodes, into the cisterna chyli, the thoracic duct, and to the left subclavian vein¹⁹ (Fig. 13-8). Most of the lower leg drains via the femoral trunks, which run on the anterior thigh to the inguinal nodes, also draining the medial and lateral thigh lymphotomes. There is a small posterior lower leg lymphotome draining to the popliteal nodes by way of the dorsolateral trunks.

A midline watershed divides the head, neck, and face areas. The right side drains to the right cervical nodes and then to the right supraclavicular nodes; the left side drains to the left cervical nodes and then to the left supraclavicular nodes. The posterior aspect of the head and neck drains into the vertebral lymphatics that drain into the supraclavicular nodes on the ipsilateral side.^{19,41}

SPECIAL IMPLICATIONS FOR THE THERAPIST 13-1

Anatomy of the Lymphatic System

It is important to realize that the right upper extremity and thoracic lymphotome drain into the right lymphatic duct and that the left upper extremity, left thoracic lymphotome, and both lower extremities, external genital areas, and abdominal lymphotomes drain into the left subclavian vein via the thoracic duct. Lymphatic obstruction or impairment affects the trunk quadrants and extremities. In addition to extremity edema, individuals may develop lymphedema of the breast, lateral trunk, abdomen, genitals, suprapubic area, or buttocks.

Drainage can be changed from one lymphotome to another by expelling lymph from an overloaded one toward a normal one, even across two or three intermediate overloaded areas (see Fig. 13-5). This change in flow occurs through the most superficial plexus, which then drains into the deeper (but still very superficial) collectors and deep trunks. The deep trunks also have collaterals crossing the watersheds to accomplish this flow. It is the dilatation of the collectors and collaterals together with the superficial plexus that accounts for the success of the therapy intervention using lymphatic drainage as part of the program.

Improper treatment of extremity edema without considering the impact of that treatment on the trunk quadrant adjacent to the limb or limbs involved can result in the development of truncal or genital edema, when none existed before intervention for the extremity edema.¹³

Lymph Nodes

Normal, healthy lymph nodes are soft and nonpalpable. Palpable lymph nodes do not always indicate serious or ongoing disease, but this determination requires an evaluation by a physician. Therapists may identify suspicious palpable lumps in a client who has already been examined by a physician. However, the therapy profession offers greater opportunity for identification of suspicious nodes, given the specificity of palpative skills and techniques practiced by a therapist. For this reason, the therapist should not hesitate to return a client to the referring or primary physician for further evaluation.

Past medical history is extremely helpful in determining the urgency of referral. A suspicious, palpable node in the presence of a previous history of cancer warrants immediate medical referral. Supraclavicular and inguinal nodes are common metastatic sites for cancer. Nodes involved with metastatic cancer are usually hard and fixed to the underlying tissue.

In acute infections, nodes are tender asymmetrically, enlarged, and matted together, and the overlying skin may be red and warm (erythematous). Changes in size (greater than 1 cm), shape (matted together), and consistency (rubbery or firm) of lymph nodes in more than one area or the presence of painless enlarged lymph nodes must be reported to the physician.

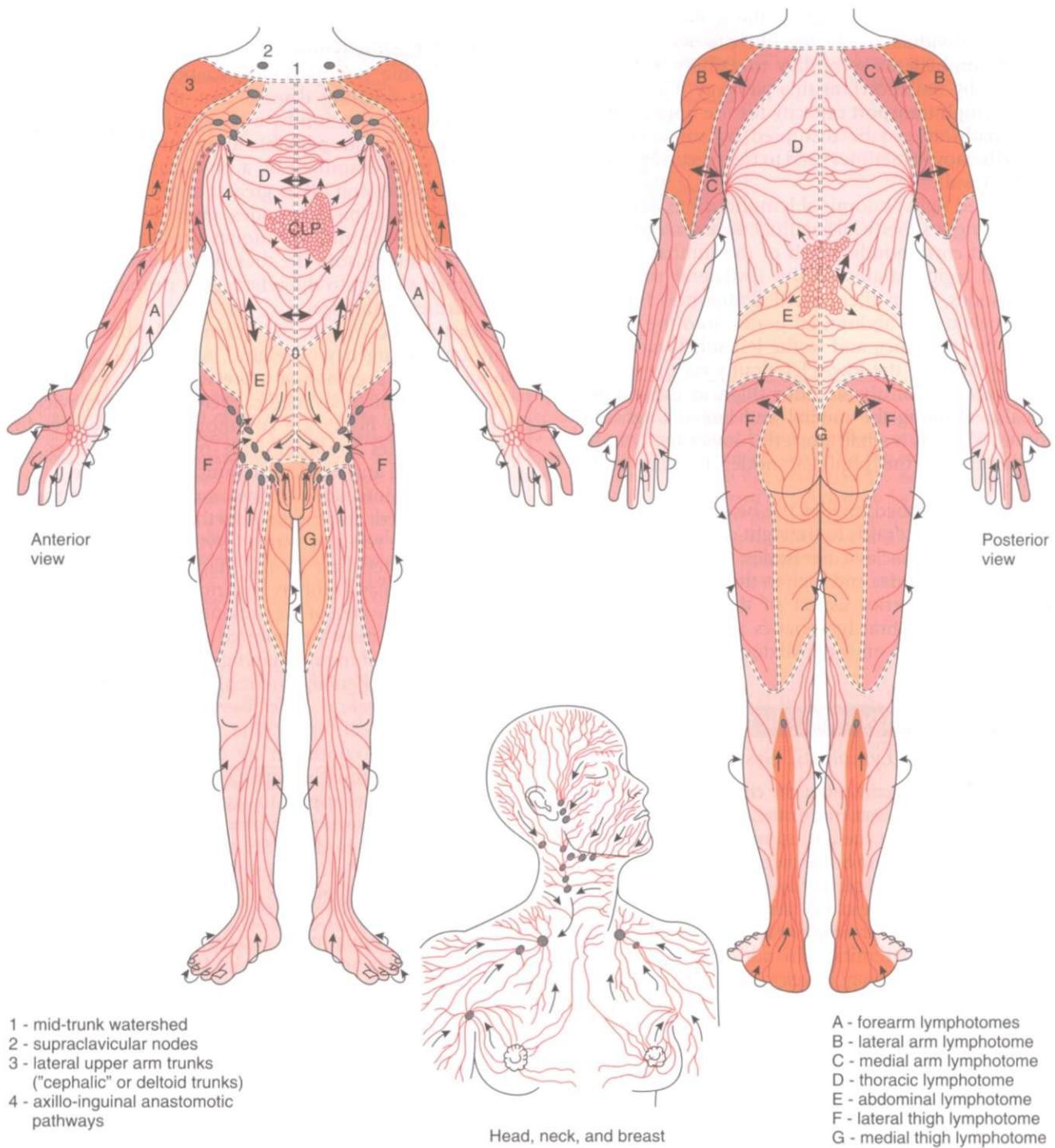
In the case of recent pharyngeal or dental infections, minor, residual enlargement of cervical nodes may be observed. Intraoral infection may also cause an inflamed cervical node. The therapist may first be alerted to this condition by a spasm of the sternocleidomastoid muscle causing neck pain.

Palpation may appear to aggravate a primary spasm, as if the spasm were originating in the muscle, when in fact a lymph node under the muscle is the source of symptoms. In such cases, the past history is the key to quickly identifying the need for medical referral or follow-up care.

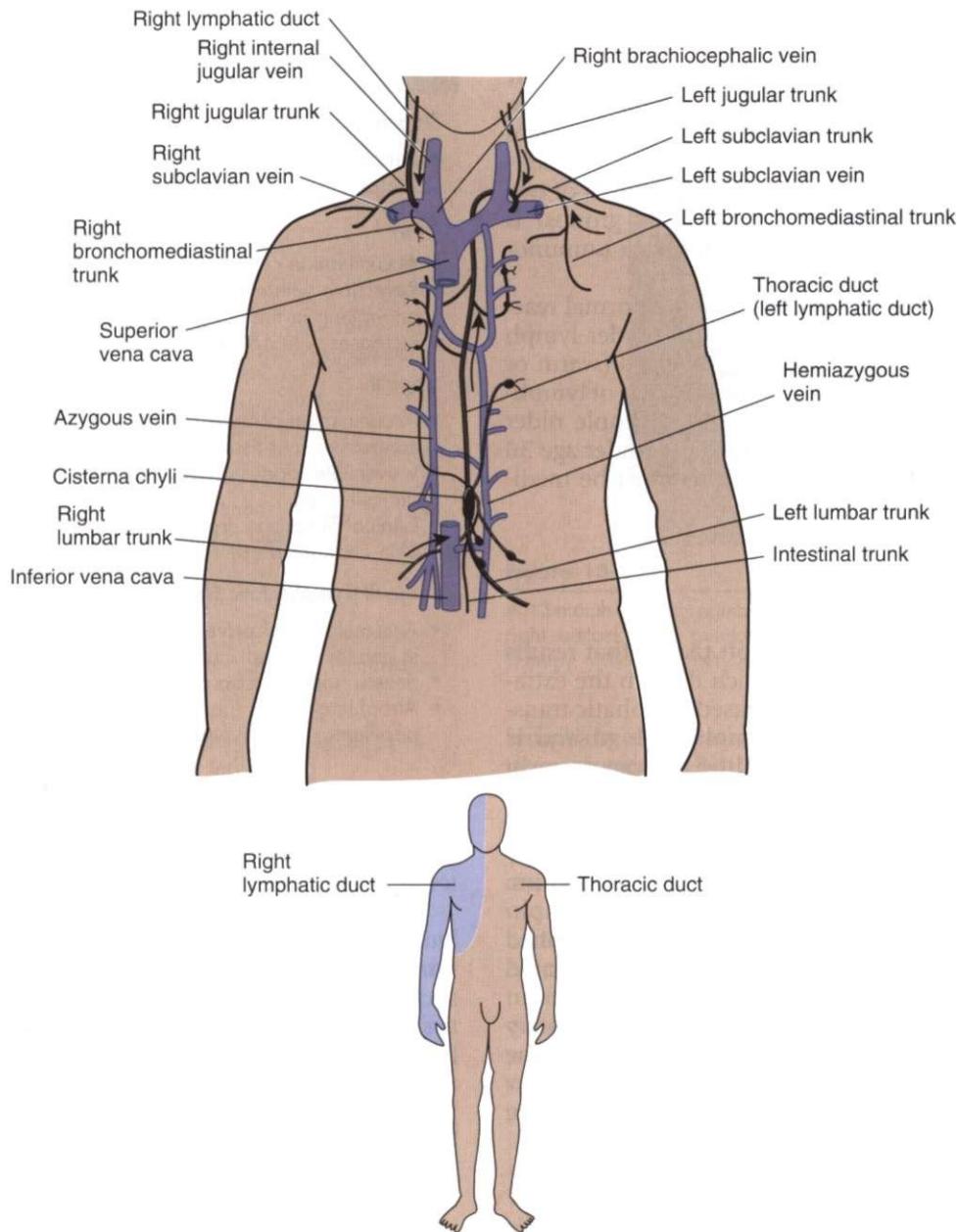
Lymphadenopathy in certain anatomic areas, such as preauricular or postauricular (in front of or behind the ear), supraclavicular, deltopectoral, and pectoral regions, is viewed by the medical community with greater suspicion because these areas are not usually enlarged as a result of local subclinical infections or trauma.⁸³

INFLAMMATION AND INFECTION IN THE LYMPHATIC SYSTEM

Disorders of the lymphatic system may result from *lymphangitis* (inflammation of a lymphatic vessel); *lymphadenitis* (inflammation of one or more lymph nodes); *lymphedema* (an increased amount of lymph fluid in the soft tissues); or *lymphadenopathy* (enlargement of the lymph nodes). Lymph nodes act as defense barriers and are secondarily involved in virtually all systemic infections and in many neoplastic disorders arising elsewhere in the body.

**Figure 13-7**

Regional lymphatic system. The dermal and subcutaneous lymph territories (lymphotomes are indicated by different shadings) of the lymphatic system are separated by watersheds marked by (— = —). Arrows indicate the direction of the lymph flow. Normal drainage is away from the watershed, but collaterals cross the watershed (thick double arrows). When the main drainage paths from each of these regions are blocked, lymph (thick single arrows) has to be carried across the watersheds via collaterals and the plexuses. The cutaneous lymphatic plexus (CLP) is shown in the center of the chest only. It is filled from the tissues and covers the entire body; this is not shown to avoid confusion. These initial lymphatics fill superficial collectors, which drain into deep ones and then into the lymphatic trunks (small arrows). The lymphotome of the external genitalia and perineum is shown but unlabeled. (From Casley-Smith JR, Casley-Smith JR: Modern treatment for lymphoedema, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia. Modified from Földi M, Kubik S: Lehrbuch der lymphologie für mediziner und physiotherapeuten mit anhang: praktische linweise für die physiotherapeute, Stuttgart, Germany 1989, Gustav Fischer Verlag.)

**Figure 13-8**

lymphatic ducts. The thoracic duct (black), leading from the cisterna chyli to discharge into the left subclavian vein, in the neck. (The blood vessels are shaded blue.) The right lymphatic duct is also shown (see figure on bottom). This carries far less lymph than the thoracic duct, draining mainly the right arm and head, the heart and lungs, and the anterior chest wall. These two main trunks sometimes are linked by large collateral lymphatics. (From Casley-Smith JR, Casley-Smith JR: *Modern treatment for lymphoedema*, ed 5, Adelaide, Australia, 1997, Lymphoedema Association of Australia. Inset, Jarvis C: *Physical examination and health assessment*, ed 3, Philadelphia, 2000, WB Saunders.)

The specific node, or nodes, affected in an infectious disease depends on the location of the infection, the nature of the invading organism, and the severity of the disease. For example, infections involving the pharynx, salivary glands, and scalp often cause tender enlargement of the neck nodes, referred to as *reactive cervical lymphadenopathy*. *Generalized lymphadenopathy*, enlargement of two or three regionally separated lymph node groups, is usually a result of inflammation, neoplasm, or immunologic reactions.

These two types of lymphadenopathy are normal reactions to infection that result in large and tender lymph nodes, but the node is not necessarily infected (warm or reddened, as with lymphadenitis). The presence of lymphadenopathy is usually more significant in people older than 50 years; lymphadenopathy in people under age 30 is usually due to benign causes, but this must be medically determined.

Lymphedema

Definition

Lymphedema is a swelling of the soft tissues that results from the accumulation of protein-rich fluid in the extracellular spaces. It is caused by decreased lymphatic transport capacity and/or increased lymphatic load and is most commonly seen in the extremities but can occur in the head, neck, abdomen, and genitalia.

Classification of Lymphedema

Lymphedema is divided into two broad categories: primary (idiopathic) and secondary (acquired) lymphedema. In the past, primary lymphedema was classified as *connatal*, if it appeared at birth; *praecox* if it appeared at puberty; or *tarda* if it developed after age 35. The term *connatal* (present from birth) applies to most primary lymphedemas that are present at birth, rather than the term *congenital*, which implies a specific genetic abnormality. The severity of lymphedema is graded using the scale from the International Society of Lymphology (ISL): Stage 0 or latent lymphedema, Stage I, Stage II, and lymphostatic elephantiasis Stage III^{130,33} (Box 13-1).

In *Stage 0* or *latent lymphedema*, lymph transport is impaired, but there is no clinical evidence of swelling. Stage 0 may last months or years before any obvious lymphedema occurs. Understanding the concept of latent lymphedema is critical in providing guidance to individuals at risk, as well as in recognizing the early signs and symptoms of progression from Stage 0 to Stage 1. These may include a sensation of heaviness, fatigue, ache, or pain in the limb at risk.

Stage I lymphedema is soft, pits on pressure, and reverses with elevation. In the early stages, there is a chronic inflammatory response to the excessive protein in the interstitium. The subcutaneous tissues begin to fibrose, progressing the lymphedema from Stage I to II. In fact, a lymphedematous limb may be Stage II in the foot and ankle and Stage I in the thigh.

Stage II lymphedema is nonpitting and does not reduce on elevation of the limb, and clinical fibrosis is present. Skin changes, such as eczema, warts, papillomas, and lymph fistulae, are common in severe Stage II lymph-

Box 13-1

STAGES OF LYMPHEDEMA

Stage 0 (Latent Lymphedema)

- Lymph transport capacity is reduced; no clinical edema is present.

Stage I

- Accumulation of protein-rich, pitting edema.
- Reversible with elevation; area affected may be normal size on waking in the morning.
- Increases with activity, heat, and humidity.

Stage II

- Accumulation of protein-rich, nonpitting edema with connective scar tissue.
- Irreversible; does not resolve overnight; increasingly more difficult to pit.
- Clinical fibrosis is present.
- Skin changes present in severe Stage II.

Stage III (Lymphostatic Elephantiasis)

- Accumulation of protein-rich edema with significant increase in connective and scar tissue.
- Severe nonpitting fibrotic edema.
- Atrophic changes (hardening of dermal tissue, skin folds, skin papillomas, and hyperkeratosis)

edema. Chronic inflammation can lead to recurrent bacterial and fungal infections.

The most severe, *Stage III lymphedema*, is referred to as lymphostatic elephantiasis. This is characterized by severe nonpitting, fibrotic edema with atrophic skin changes such as thickened, leathery, keratotic skin, skin folds with tissue flaps, warty protrusions, papillomas, and leaking lymph fistulae. Lymphangiomas (form of lymphangiectasia) may also be present.

Incidence

The exact incidence of primary lymphedema is unknown because many people remain undiagnosed or if diagnosed, treatment or follow-up care does not occur, and the condition remains unreported.³³ Approximately 15% of primary lymphedemas are present at birth (formerly called *connatal*). The most common form of primary lymphedema occurs from adolescence to midlife and accounts for 75% of primary lymphedema in a 4:1 ratio of females to males (formerly called *lymphedema praecox*). Of all primary lymphedema, 10% to 20% appears abruptly after age 35 (formerly called *lymphedema tarda*).³³ A small percentage of primary lymphedemas occur in association with rare genetic syndromes, such as Milroy's (appears at birth) and Meige's (develops anywhere from early childhood to puberty) diseases, accounting for approximately 2% of primary lymphedemas.

The incidence of secondary lymphedema also remains an approximate figure. Lymphatic filariasis affects over 120 million people in 80 countries throughout the tropics and subtropics of Asia, Africa, the Western Pacific, and parts of the Caribbean and South America. Presently, no detailed maps are available of the geographical