Arteries are classified into three categories on the basis of their size and histologic features: (1) large or elastic arteries (the aorta and its immediate proximal, larger branches, including the innominate, subclavian, common carotid, and pulmonary arteries); (2) medium-sized or muscular arteries (located just distal to elastic arteries, including the common femoral, axillary, and carotid arteries); and (3) small arteries (usually <2 mm in diameter) that course in the substance of tissues and organs. This chapter considers diseases of medium and small arteries.

Arterial Anatomy

All arteries possess three layers: the tunica intima, tunica media, and tunica adventitia. As peripheral arteries diminish in caliber, these three layers become progressively indistinct and are no longer identifiable at the level of the arteriole (precapillary vessel containing smooth muscle).

The tunica intima has an inner lining of endothelial cells surrounded by subendothelial connective tissue. The single layer of continuous endothelium is a unique thromboresistant layer between blood and the potentially thrombogenic subendothelial tissues. The integrity of the endothelium is a fundamental requirement for maintenance of normal structure and function of the entire vessel wall. Endothelial injury can result in intraluminal thrombosis and may contribute to the initiation of atherosclerosis.

The tunica media is made up primarily of circular or spiral smooth muscle cells arranged in concentric layers. The outer limit of this layer is marked by a well-defined, external elastic membrane. The elastic content of the tunica media gives resilience to medium-sized arteries. With age, the elastic fibers deteriorate and are replaced by fibrous tissue. This loss of elasticity results in stretching and elongation and accounts for the progressive tortuosity and development of arterial aneurysms with aging. Vascular smooth muscle cells may be important in lipid accumulation in the vessel wall during atherosclerosis and participate in vasoconstriction and dilation.

The tunica adventitia is a poorly defined layer of connective tissue in which nerve fibers and small, thin-walled nutrient vessels (vasa vasorum) are dispersed. Medium-sized arteries contain more nerve fibers than larger vessels, reflecting the importance of their role in the autonomic regulation of blood flow.

The peripheral arterial vascular system can be considered as a single end-organ subject to eight basic pathophysiologic processes: (1) atherosclerosis, (2) aneurysm, (3) embolism, (4) thrombosis, (5) inflammation, (6) trauma, (7) vasospasm, and (8) arteriovenous fistula. Two of these—atherosclerosis and thrombosis—are responsible for most peripheral arterial problems.

Pathophysiology

Atherosclerosis

Atherosclerosis is a disease of large- and medium-sized muscular arteries. The basic lesion, the atheroma, or fibrofatty plaque, is a raised focal plaque within the intima; it has a lipid core (mainly cholesterol, usually complexed to proteins and cholesterol esters) covered by a fibrous cap. As the plaques increase in size and number, they progressively encroach on the lumen of the artery and the adjacent media. Atheromas compromise arterial blood flow and weaken the walls of the affected arteries.

The distribution of atherosclerotic plaques is rather constant. The abdominal aorta has more atherosclerotic disease than the thoracic aorta, and aortic lesions are much more common and prominent around the ostia of major branches. Other vessels affected by atherosclerosis are the aortoiliac, femoral, and popliteal arteries; the descending thoracic aorta; the coronary arteries; and the circle of Willis. Vessels of the upper extremities are usually spared.

As atherosclerosis progresses, atheromas almost always calcify, resulting in hard, brittle vessels. Ulceration of the luminal surface and rupture of the atheromatous plaques discharge debris, producing atheroemboli (cholesterol emboli). Fissured or ulcerated lesions can produce in situ thrombosis, causing acute intraluminal occlusion.

Hemorrhage into the plaque may further compromise the arterial lumen. Although atherosclerosis primarily affects the intima, in severe cases, the tunica media undergoes pressure atrophy and loss of elastic tissue, with sufficient weakening to create aneurysmal dilation.

Aneurysms

A true aneurysm is an abnormal localized dilation of the intact vessel wall. With a pseudoaneurysm the entire wall perforates or ruptures, and the extravasated blood is contained by the surrounding tissues, eventually forming a fibrous sac that communicates with the artery.

Mural and mechanical factors contribute to true aneurysm formation. The major cause of aneurysms is a weakness or defect in the integrity of the arterial wall. The only aneurysms that develop in a normal arterial segment are poststenotic aneurysms, such as with coarctation. Acceleration of flow past a narrow point creates slower flow beyond the stenosis lateral to the jet stream, producing
increased lateral pressure. Aneurysmal dilation accelerates, increasing the risk of rupture as diameter increases, as predicted by Laplace’s law: tension (lateral pressure) in the wall of a hollow viscus varies directly with its radius (tension = pressure × radius).

The most common cause of aneurysms is severe atherosclerosis resulting from thinning and destruction of the tunica media. Atheromatous ulcer covered by mural thrombi are common within an aneurysm. Such mural thrombi can form emboli that lodge in distal vessels. When an entire aneurysm is filled with thrombus material, arterial occlusion results.

Aneurysms cause clinical symptoms through (1) rupture with subsequent hemorrhage, (2) impingement on adjacent structures, (3) occlusion of a vessel by either direct pressure or mural thrombus formation, (4) embolism from mural thrombus, and (5) presentation as a pulsatile mass.

Arterial Embolism
An embolus is a blood clot or other foreign body that is carried by the blood to a site distant from its point of origin. Most emboli are the result of detached thrombus formation (thromboembolism). Less common sources include debris from ruptured atherosclerotic plaques, tumor debris, or foreign bodies. Unless otherwise specified, the term embolus in this chapter is defined as thromboembolus.

Thromboembolism. Most arterial emboli (85%) originate in thrombus formation in the heart. Left ventricular thrombus formation resulting from myocardial infarction accounts for 60 to 70% of arterial emboli. Atrial thrombi associated with mitral stenosis and rheumatic heart disease account for only 5 to 10% of arterial emboli.2 Coexisting atrial fibrillation, often without mitral stenosis, is present in 60 to 75% of patients with peripheral arterial embolic events, because atrial fibrillation itself can predispose patients to intracardiac clotting.3

Acute arterial emboli often cause distal tissue infarction. Clinical outcome depends mostly on the amount of collateral circulation present but also on the size of the vessel and the degree of obstruction. Patients with long-standing atherosclerosis have well-developed collateral circulation, whereas sudden occlusion of a normal artery without collateral pathways results in severe ischemia. After acute obstruction, the embolus can propagate proximally or distally, fragment and embolize further to distal vessels, or precipitate associated venous thrombosis by initiating a localized inflammatory reaction.

Because vessel diameters change most abruptly at branch points, embolic occlusion most often occurs at major arterial bifurcations. The bifurcation of the common femoral artery is the most frequent site of arterial embolism, accounting for 35 to 50% of all cases.2 The smaller femoral and popliteal arteries are involved twice as often as the larger aortic and iliac vessels, reflecting the small size of most emboli.

Cell death from arterial ischemia can produce high concentrations of potassium, lactic acid, and myoglobin in the extremity distal to an arterial occlusion. Their sudden release after revascularization can produce life-threatening hyperkalemia, metabolic acidosis, and myoglobinuria. This myonephropathic-metabolic syndrome accounts for approximately one third of the deaths from arterial embolism after revascularization.4

Atheroembolism. Atheroembolism refers to microemboli consisting of cholesterol, calcium, and platelet aggregates dislodged from proximal complicated atherosclerotic plaques that lodge in distal end arteries. In the central nervous system, atheroemboli cause transient ischemic attacks and strokes. In the peripheral vascular system, atheroemboli characteristically cause cool, painful, and cyanotic toes, or the blue toe syndrome.7 Atheroemboli are caused by a proximally located arterial lesion, usually atherosclerotic plaques or aneurysms. Bilateral distal extremity involvement usually implies an aortic source, whereas unilateral atheroemboli usually arise from sites distal to the aorta. Distal lesions are most common in the femoropopliteal arteries (60%) and the aortoiliac arteries (40%). Aortic lesions (e.g., aneurysms, polytetrafluoroethylene grafts) are a less common source of microemboli.5

Atheroemboli are small and tend to lodge in distal arteries, such as the digital arteries, which are 100 to 200 µm in size. Single atheroembolic events seldom result in tissue loss, but atheroemboli tend to cluster. If unrecognized, repeated events ultimately result in loss of collateral circulation, progressive symptoms, and extensive tissue infarction.5

Infectious emboli from bacterial endocarditis can produce septic infarcts that may convert to large abscesses. Rarely, cardiac and noncardiac tumors or foreign bodies may gain access to the arterial circulation and embolize. Primary or metastatic lung neoplasms, malignant melanoma, and bullet emboli have been reported. In patients with cyanotic congenital heart disease (e.g., patent foramen ovale), venous emboli may pass directly to the arterial circulation (paradoxical emboli). Although rare, this possibility should be considered in any patient with simultaneous arterial and venous emboli, particularly if a source of the arterial embolus is not evident.

Arterial Thrombosis
Thrombosis is the in situ formation of a blood clot within the uninterrupted arterial vascular system. Complicated atherosclerotic plaques are usually responsible for the two major factors that cause in situ thrombosis: endothelial injury and alterations in normal blood flow. Less common causes include acute vasculitis and trauma. Thrombosis is rare in normal arteries.5

Peripheral arterial thrombi are usually occlusive, although they may be limited to one wall (mural) in larger vessels. Peripheral arterial thrombi are usually firmly attached to the damaged arterial wall and infrequently embolize. The clot may propagate proximally and distally, which intensifies ischemia.

Inflammation
Inflammatory arterial injury can be caused by drugs, irradiation, mechanical trauma, or bacterial invasion. The major cause of arteritis is noninfectious systemic necrotizing vasculitis. Most cases of infectious arteritis are caused by direct invasion of the arterial wall. Septicemia, intravenous drug abuse, or infective endocarditis is most often responsible. Certain fungal infections, particularly aspergillosis and mucormycosis, are frequently associated with vasculitis and thrombosis.

Trauma
Different types of vascular injury result in characteristic pathologic syndromes. Partial arterial lacerations continue to bleed because the intact portion of the vessel wall prevents retraction and closure of the arterial wound. This may form an expanding hematoma, causing progressive deformity, pain, and nerve compression. Complete arterial transection usually has only moderate or insignificant bleeding because of arterial spasm of the transected ends of the artery and the formation of a temporary thrombus. Delayed hemorrhage in completely transected arteries may result from relaxation of arterial spasm, eventual liquefaction of the thrombus, or displacement of the thrombus by arterial pressure. Blunt injury may produce partial or complete intimal disruption. Dissection of the distal intima can lead to progressive obstruction and thrombosis. Complete occlusion may not occur for hours or days after injury. Vasospasm can accompany injuries that are adjacent to blood vessels; spontaneous
resolution always occurs in the absence of arterial disruption or intimal injury.

Vasospasm

Vasospastic disorders (Raynaud’s disease, Raynaud’s phenomenon, livedo reticularis, acrocyanosis, erythromelalgia) produce an abnormal vasomotor response in distal small arteries. The cause of these disorders is unknown but may be related to the autonomic innervation of the peripheral arterioles. The vasospastic disorders are characterized by the presence of ischemic symptoms and the absence of tissue loss. True organic changes within the arterial wall are absent. In contrast, patients with digital ulceration and gangrene always have fixed arterial occlusions in the distal extremity arteries.

Arteriovenous Fistulae

Abnormal communication between arteries and veins may result from congenital defects, rupture of an arterial aneurysm into an adjacent vein, penetrating injuries, and inflammatory necrosis associated with neoplasms or infection. Arteriovenous fistulae can occur in any region of the body. The artery proximal to the fistula becomes distended, tortuous, and aneurysmal. Similar changes occur in the venous side of the fistula. Proximal and distal veins respond to alterations in hemodynamics with intimal proliferation and fibrosis, followed by a decrease in the internal elastic lamina, resulting in distention, tortuosity, and aneurysm formation. The resultant chronic venous hypertension may cause dermatitis and ulceration of overlying skin. The size of the fistula generally increases with time.

Approximately 60% of arteriovenous fistulae are associated with a false aneurysm. False aneurysm formation can occur as part of the fistulous tract or as the result of arterial or venous dilation.

The increase in cardiac output that occurs when blood switches from the arterial to the venous system can result in a widened pulse pressure or high-output cardiac failure.

CLINICAL FEATURES

History

Patients with peripheral arterial disease have pain, tissue loss (ulceration or gangrene), or a change in sensation or appearance (swelling, discoloration, or temperature change). Because the primary cause of peripheral arterial disease is atherosclerosis, related conditions providing evidence of atherosclerosis are cardiac disease, myocardial infarction, cardiac dysrhythmias (e.g., atrial fibrillation), stroke, transient ischemic attacks, and renal disease. Factors that increase the likelihood of atherosclerosis are cigarette smoking, diabetes, hypercholesterolemia, and hypertension. Intravenous drug use can lead to arterial injury.

Risk factors not related to atherosclerosis include prior injuries or surgeries, major illnesses, a history of phlebitis or pulmonary embolism, the presence of autoimmune disease or arthritis, and a history of prior coagulation abnormalities.

Acute Arterial Occlusion

The patient with acute arterial occlusion usually exhibits some variant of the five Ps: pain, pallor, pulselessness, paresthesias, and paralysis. Paresthesias and paralysis indicate limb-threatening ischemia that requires emergency surgical intervention regardless of the cause. In patients with non-limb-threatening ischemia, accurate differentiation between embolism and in situ thrombosis as the cause of acute arterial occlusion determines management. Arterial embolism is best managed by emergency Fogarty catheter embolectomy. Non-limb-threatening ischemia from in situ thrombosis is often aggravated by emergency surgical intervention and is therefore initially best managed nonoperatively, if possible (Fig. 87-1). Because acute arterial embolism usually occurs in patients without significant peripheral atherosclerosis and without well-developed collateral circulation, it usually manifests as sudden limb-threatening ischemia. Patients describe a sensation of the leg’s being “struck” by a severe shocking pain. Often the patient has to sit or fall to the ground during the sudden event.

In situ thrombosis usually occurs in patients who have longstanding significant peripheral atherosclerosis and well-developed collateral circulation and is often seen subacutely with non-limb-threatening ischemia. A history of claudication is common with in situ thrombosis and rare in patients with arterial embolism.

Chronic Arterial Insufficiency

Chronic arterial insufficiency causes two characteristic types of pain: intermittent claudication and ischemic pain at rest. The location of arterial occlusion determines the location of claudication. Calf claudication is associated with femoral and popliteal disease, typically a cramping pain, reliably reproduced by the same degree of exercise and completely relieved by rest (usually 1-5 minutes). Aortoiliac occlusive disease causes claudication in the buttocks and hips, as well as the calves. The calf pain in aortoiliac disease is generally more severe than the buttock and thigh pain, which is more often described as an aching, discomfort, or weakness. Some patients deny pain, complaining only that the thigh or hip “gives out” with exercise. Aortoiliac occlusive disease severe enough to produce bilateral claudication is almost always associated with impotence in men (Leriche’s syndrome). Even in the absence of impotency, bilateral hip or thigh pain in a man should indicate the possibility of aortoiliac occlusive disease.
Chronic arterial insufficiency may progress so that ischemic pain occurs at rest. Rest pain often begins in the feet and typically involves the foot distal to the metatarsals, awakening the patient from sleep. Ischemic rest pain is a severe, unrelenting pain aggravated by elevation and unrelieved by analgesics, but patients have prompt relief with any activity that involves a standing position. Patients often sleep with the leg dangling over the side of the bed or sleep in a chair to improve perfusion pressure to the distal tissues.

**Physical Examination**

A systematic assessment of the peripheral vascular system includes palpation of the pulse volume in the pairs of brachial, radial, femoral, posterior tibial, and dorsalis pedis arteries documented on a scale of 0 to 4+. Approximately 10% of the population does not have one of the dorsalis pedis pulses. Carotid arteries should be gently palpated one at a time. The lower extremities should be examined for signs of chronic and advanced ischemia. Muscular atrophy, particularly in the lower extremities, and loss of hair over the toes and feet with thickening of the toenails resulting from slowness of nail growth are common signs of arterial insufficiency. As ischemia becomes more advanced, the skin becomes shiny, scaly, and “skeletonized” from atrophy of the skin, subcutaneous tissue, and muscle.

Areas where ischemia is suspected can be tested by blanching with finger pressure; a delay in return of normal color (compared with that of the unaffected extremity) implies reduced perfusion.

Buerger’s sign provides reliable evidence of severe advanced ischemia. With the patient supine, his or her legs are elevated to 45 degrees to bring the feet more than 12 inches above the right atrium, and any pallor of the feet is noted. If the color does not change, the patient dorsiflexes the feet five or six times; pallor induced by exercise also connotes inadequate arterial flow. The patient is then moved to the sitting position with the feet hanging down. Within 10 to 15 seconds, color should return, and the veins should fill. Typically in the ischemic foot the first color return is cyanotic, transitioning to red as reactive hyperemia occurs. If the veins require more than 20 seconds to become distended, advanced ischemia is present. With severely restricted arterial inflow and chronic dilation of the peripheral vascular bed, the foot turns chalk white on elevation and intensely hyperemic after 1 minute of dependency. Localized pallor or cyanosis associated with poor capillary filling is usually a prelude to ischemic gangrene or ulceration.

Doppler ultrasonography should be used in patients with questionable or absent pulses. The ankle-brachial index (ABI) is made by comparing the systolic blood pressure at the level of the ankle with the brachial systolic pressure. With the patient supine, a blood pressure cuff is applied just proximal to the malleolus, inflated above brachial systolic pressure, and then deflated slowly. Ankle systolic pressure can be accurately measured with a Doppler probe placed over the dorsalis pedis or posterior tibial artery. This pressure is normally 90% or more of the brachial systolic pressure; with mild arterial insufficiency, it is between 70 and 90%; with moderate insufficiency, between 50 and 70%; and with severe insufficiency, less than 50%.

The Allen test is helpful in assessing patency of the radial or ulnar artery distal to the wrist. The patient initially opens and closes the hand and then clenches the fist to expel as much blood from the hand as possible; the examiner then compresses the radial and ulnar arteries. When the patient opens the fist, the hand is pale. The examiner then releases pressure from the radial artery but maintains it on the ulnar artery. If the radial artery distal to the wrist is patent, the hand becomes pink rapidly; if it is occluded, the hand remains pale. The maneuver is then repeated by maintaining pressure on the radial artery while releasing the ulnar artery. A comparison can be made with the opposite hand.

### Arterial Embolism

The physical examination can differentiate arterial embolism from in situ thrombosis. The sudden loss of a pulse is the hallmark of arterial embolism but may be difficult to recognize if the prior pulse status is unknown or is abnormal as the result of associated atherosclerosis. A bounding pulse may be felt initially at the location of an embolus from transmitted pulsations through the fresh clot. In general, patients with arterial embolism have few physical findings suggestive of long-standing peripheral vascular disease with normal proximal and contralateral limb pulses. Tenderness to palpation may occur at the site of an embolic occlusion.

If arterial embolism is suspected, the physical examination should be directed toward identifying its source, most commonly a left ventricular mural thrombus secondary to a prior myocardial infarction and a left atrial thrombus in a patient with mitral valve disease. Coexistent atrial fibrillation is common.

The limb distal to an embolic occlusion is initially chalk white. Because of absence of blood from the venules of the subcapillary layer, the demarcation between ischemic and nonischemic tissue is sharp. With time, cyanosis may appear, indicating desaturation of blood with continued ongoing ischemia. Paresthesia or paralysis indicates limb-threatening ischemia. The presence of sensitivity to light touch is often the best guide to viability of the tissue. Complete anesthesia demands immediate surgical intervention. Paralysis represents severe skeletal muscle and neural ischemia, which may be irreversible. Involuntary muscle contracture with woody hardness represents irreversible ischemia.

### Arterial Thrombosis

Physical findings of in situ thrombosis are often accompanied by evidence of atherosclerotic occlusive disease. Proximal or contralateral limb pulses are usually diminished or absent. An embolic source, such as mitral valve disease or atrial fibrillation, is usually absent. Because of collateral circulation, demarcation of limb ischemia is less well defined in these patients (Table 87-1).

Carotid, renal, and femoral arteries may have bruits, and there may be an abdominal aortic aneurysm. If an occlusion of the

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<th><strong>Table 87-1</strong> Differentiation of Embolus from Thrombosis</th>
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<td><strong>CLINICAL FINDINGS</strong></td>
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<td>Identifiable source for embolus</td>
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<td>History of claudication</td>
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<td>Physical findings suggestive of occlusive disease</td>
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upper extremity vessels is suggested, the subclavian artery should be evaluated by palpating for thrills and listening for bruits in the supraclavicular fossa.

A funduscopic examination may yield evidence of arteriosclerosis or hypertension. Hollenhorst plaques (atheromatous emboli containing cholesterol crystals in the retinal arterioles) may be detected. Roth’s spots (round or oval white spots seen near the optic disk) may be present in patients with infective endocarditis.

Embolic phenomena can cause diverse end-organ damage: hemiplegia from cerebral emboli, flank pain with hematuria from renal emboli, left upper quadrant abdominal pain from splenic infarcts, and pleuritic pain with hemoptysis from pulmonary emboli. Septic pulmonary embolism from right-sided endocarditis may be confused with pneumonia.

**Inflammation**

Inflammatory vascular disease manifests primarily as skin involvement. Skin lesions typically appear as palpable purpura; other cutaneous manifestations of vasculitis include macules, papules, vesicles, bullae, subcutaneous nodules, ulcers, and recurrent or chronic urticaria. The skin lesions may be pruritic or even painful, with a burning or stinging sensation. Lesions are more common in dependent areas: in the lower extremities in ambulatory patients or in the sacral area in bedridden patients. Edema accompanies some lesions, and hyperpigmentation occurs in areas of recurrent or chronic lesions.

**Vasospasm**

Vasospastic disorders cause a sharp border between ischemic and normal tissue. Raynaud’s disease is characterized by intermittent attacks of triphasic color changes: pallor, cyanosis, and then rubor. The most important element is pallor, during which the digits turn chalk white. Attacks last 15 to 60 minutes, and rewarming the hands restores normal color and sensation. Color changes do not occur above the metacarpophalangeal joints and rarely involve the thumb.

Livedo reticularis is characterized by a persistent cyanotic mottling of the skin that has a typical “fishnet” appearance and may involve all parts of the extremities and trunk. Acrocyanosis, the least common vasospastic disorder, is characterized by persistent, painless, diffuse cyanosis of the fingers, hands, toes, and feet. Cyanosis usually intensifies with exposure to cold and decreases with warming. The involved parts are nearly always cold, exhibit excessive perspiration, and have normal arterial pulses.

**Arteriovenous Fistulae**

Arteriovenous malformations and fistulae, although rare, must be distinguished from vascular bruits or aneurysms. True aneurysms and arterial stenoses are associated with a systolic murmur. Pseudoaneurysms generally have a loud systolic and sometimes a separate diastolic murmur. Arteriovenous fistulae have a constant systolic and diastolic (to-and-fro) murmur heard best directly over the lesion and often associated with a palpable thrill, precisely analogous to the findings of a therapeutic dialysis arteriovenous fistula. Unless congenital, arteriovenous fistulae occur at prior operative or trauma sites. The skin overlying the lesion may be warm, but distally the temperature is often decreased. Veins peripheral to the fistula are usually distended and varicose. Large and long-standing arteriovenous fistulae produce high cardiac output and widened pulse pressure. Digital pressure on the artery leading to the fistula or the fistula itself may decrease the tachycardia (Branham’s sign).

**Diagnostic Strategies**

An accurate diagnosis of peripheral arterial occlusive disease can be achieved in most patients by careful history and physical examination supplemented by bedside testing.

**Noninvasive Assessment**

Doppler ultrasonography measures blood flow velocity by detecting the frequency shift of sound waves reflected from red blood cells that move toward or away from the transducer. The Doppler signal generates a normal triphasic velocity waveform. Progressive arterial narrowing alters the triphasic waveform to biphasic and finally monophasic shape. Such Doppler ultrasonographic waveform analysis can detect significant arterial occlusive disease, although it is less accurate in determining exact location.

Ultrasonic is useful in detecting and evaluating atherosclerotic plaques and mural thrombi and in sizing aneurysms of the abdominal aorta and iliac, femoral, and popliteal arteries. B-mode ultrasonography is noninvasive, painless, less expensive than other modalities, and universally available and is the diagnostic procedure of choice for the initial evaluation and determination of the size of peripheral artery aneurysms. Bedside ultrason can lead to rapid diagnosis of life-threatening conditions and reduce the number of delayed or invasive diagnostic procedures.

B-mode duplex ultrasonography combines the image of B-mode ultrasonography and sophisticated online computer analysis of accurately sampled Doppler waveforms to allow simultaneous acquisition of both the image of a vascular structure and the characteristics of blood flow velocity within it. Duplex scanning permits noninvasive and accurate diagnosis of peripheral vascular, cerebrovascular, and venous disease.

Color imaging of blood flow combined with duplex scanning is known as color-coded Doppler, Doppler angiography, or angiodynography. The procedure of choice for most conditions, this combination allows noninvasive and accurate detection of atherosclerotic plaques and stenoses, their effect on intraluminal blood flow, and the presence of venous thrombosis.

**Contrast Arteriography**

Angiography is the definitive test of abnormal peripheral artery anatomy but is often inconclusive about the physiologic condition of the tissues. Adverse effects of contrast media and catheter-related complications must be weighed against the benefits of this procedure. Contrast media have a direct toxic effect on vascular endothelium; can produce renal failure, especially in diabetic patients; may cause peripheral vasodilation with hypotension; may result in seizures and stroke in patients with neurologic conditions; and can cause severe idiosyncratic and allergic reactions. Catheter-related complications, including embolization, catheter breakage, and vascular disruption, vary with operator skill and anatomic location but average 0.5%. The overall mortality rate from angiography is 0.03%. Emergency angiography is usually necessary in the following circumstances: (1) acute arterial embolus or thrombosis if the clinical diagnosis is uncertain, (2) consideration of emergency vascular bypass grafting, and (3) characterization of vascular abnormality before emergency surgical correction.

**Computed Tomography and Magnetic Resonance Imaging**

Computed tomography angiography is the most useful test for evaluation of the abdominal aorta. In the peripheral arteriovascular system, CT angiography is useful primarily for atherosclerotic, infected, and false aneurysms and for imaging the cerebral
circular. Magnetic resonance imaging (MRI) has the capability for angiography (magnetic resonance angiography) and has been particularly useful in delineating cerebrovascular problems (see Chapter 101); it is seeing expanded use in the evaluation of peripheral vascular disease. The ability to make axial, coronal, and sagittal sections provides accurate visualization of anatomy. MRI detects changes in the relaxation variables of tissues before obvious structural changes, uniquely differentiating blood, thrombus, fat, and fibrosis.

**MANAGEMENT OPTIONS**

The management of acute arterial occlusion depends on the degree and cause of ischemia. Patients with limb-threatening ischemia from embolism should undergo emergency Fogarty catheter embolectomy. Patients with limb-threatening ischemia caused by in situ thrombosis require direct or Fogarty catheter thrombectomy combined with vascular bypass grafting. Thrombectomy alone often fails because of recurrent thrombosis. Patients who have a lesion that cannot be bypassed, who have evidence of irreversible ischemia, or who are too ill to tolerate revascularization are treated with primary amputation.

A patient with non–limb-threatening ischemia from embolism still is treated with Fogarty catheter embolectomy. Non–limb-threatening ischemia from in situ thrombosis is managed nonoperatively with immediate systemic heparinization and possibly with intra-arterial fibrinolytic therapy (see Fig. 87-1).

Elective surgical repair of an asymptomatic atherosclerotic peripheral arterial aneurysm is usually accomplished by excision of the aneurysm with end-to-end anastomosis or graft interposition. Infected true and false peripheral aneurysms require aneurysm resection, debridement of infected tissue, and ligation of the proximal and distal uninfected arteries. Autogenous vein bypass through uninfected tissue planes is attempted, because prosthetic grafts carry a high risk of graft infection. The surgical approach for infected false aneurysms is similar to that for peripheral atherosclerotic aneurysms.

Patients with thoracic outlet syndrome who have cervical ribs, arterial involvement, or significant neurologic symptoms require surgical decompensation with removal of anomalous fibromuscular bands and resection of the first rib, if present. Subclavian and subclavian-axillary aneurysms can be treated with resection and end-to-end anastomosis, graft reconstruction, or surgical revision. Patients with distal embolic occlusions are treated with Fogarty catheter embolectomy. Axillary and subclavian vein thromboses are best managed with surgical thrombectomy or systemic fibrinolytic therapy. Patients with only brachial plexus involvement and minimal signs and symptoms should be followed closely with conservative treatment.

Surgical treatment of peripheral arteriovenous fistulae requires interrupting the fistula tract and restoring both arterial and venous continuity with end-to-end anastomosis or graft interposition. If the anatomic location precludes surgical intervention, percutaneous transvascular embolization with liquid tissue adhesives (e.g., isobutyl 2-cyanoacrylate) is usually successful.

**Noninvasive Therapy**

**Acute Anticoagulation with Heparin**

For acute arterial embolism, acute arterial thrombosis, and subclavian vein thrombosis, heparin is indicated at standard intravenous doses (80 units/kg by intravenous bolus, followed by a maintenance infusion of 18 units/kg/hr). Heparin quickly reduces thrombin generation and fibrin formation, minimizing clot propagation, which can intensify limb ischemia and jeopardize tissues. Relative contraindications include recent neurosurgery (especially within 2 weeks), major surgery within 48 hours, childbirth within 24 hours, a known bleeding diathesis, thrombocytopenia, a potentially hemorrhagic lesion, and active bleeding.

**Fibrinolytic Therapy**

Low-dose intra-arterial fibrinolytic therapy is increasingly used for acute arterial occlusion. Patients with limb-threatening ischemia are not candidates because clot lysis generally takes 6 to 72 hours. Patients with limb-threatening ischemia cannot tolerate several more hours of ischemia without tissue or limb loss. Fibrinolytic therapy is generally reserved for patients with in situ thrombosis and non–limb-threatening ischemia.

Intra-arterial fibrinolytic agents induce clot lysis in the small, distal runoff vessels, decreasing outflow resistance and enabling the native artery to remain open longer. Fibrinolysis often uncovers a critical stenosis that, untreated, may lead to another episode of thrombosis. After successful fibrinolytic therapy, most patients require secondary bypass grafting or percutaneous transluminal angioplasty. Streptokinase, urokinase, and tissue plasminogen activator have all been used successfully. Intravenous administration of a fibrinolytic agent is less effective than direct administration into the clot. Clots more than 30 days old are more organized and less likely to achieve successful lysis.

**Invasive Therapy**

**Fogarty Catheter Thrombectomy**

The Fogarty catheter is most frequently used for iliac, femoral, and popliteal embolectomy, often with only local anesthesia. Aortic saddle embolus is removed by sequentially passing the Fogarty catheter through bilateral common femoral arteriotoomies. Newly formed in situ thrombosis can often be successfully removed with the Fogarty catheter. An older thrombus adheres more firmly to the damaged vessel wall, requiring direct surgical thrombectomy. The Fogarty catheter is not used in the venous system because of valves.

**Peripheral Percutaneous Transluminal Angioplasty**

The initial success and long-term patency achieved with angioplasty depend on the location of the lesion and the extent of atheromatous disease. Proximal larger arteries (e.g., iliac, femoral-popliteal) have the best initial and long-term results. Discrete stenotic lesions (<5 cm) have better long-term patency rates than vessels that are diffusely involved or have multiple involved segments. Balloon angioplasty is the accepted treatment for isolated stenotic lesions in the renal, iliac, and superficial femoral vessels.

Transluminal angioplasty with intravascular stent is used in more distal vessels, including the popliteal and tibial circulation, in cases of more diffuse lesions, and for patients who are prohibitive surgical risks, although its value remains to be determined.

Recanalization devices include the percutaneous atherectomy catheter, percutaneous angioscope, hot-tip laser, excimer laser, and high-speed rotating wire and drill.

**Grafting**

Vascular grafting is associated with a variety of complications that can be diagnosed in the emergency department. Autogenous vein grafts (usually a reversed greater saphenous vein) provide excellent long-term patency for small arteries. Vein grafts respond to arterial pressure with gradual intimal proliferation and medial fibrosis. They may develop atherosclerosis, which can lead to graft stenosis and thrombosis. False aneurysms can form along the suture line.
Polytetrafluoroethylene (Teflon) prosthetic grafts are widely used in medium and large arteries that are impossible to bridge with smaller vein grafts. Prosthetic grafts have a higher rate of thrombosis than venous grafts. Distal emboli may result from poor fixation of luminal fibrin. If the prosthetic graft has not been adequately covered by viable tissue, it can erode into adjacent structures and hollow viscera. Prosthetic graft infection is a devastating complication requiring removal of the entire graft.

Vascular grafts can be used to bypass arterial occlusions and reconstruct a diseased arterial bifurcation, or can be interposed between sections of resected artery. The two most common complications of both prosthetic and vein grafts are thrombosis and development of a false aneurysm at one or more suture lines. Bypass grafting is most often used as palliative treatment for symptoms of atherosclerotic occlusive disease. Patients with localized unilateral stenosis (<3-5 cm in length) may have a comparable rate of success with percutaneous transluminal angioplasty with or without stent placement.\(^\text{15}\)

Patients with calf claudication from superficial femoral or popliteal occlusive disease can slow progression if they stop smoking and maintain an active exercise regimen. Patients who have progression of disease, significant rest pain, or tissue loss require surgical revascularization.

**Sympathectomy**

Lumbar sympathectomy is no longer used for treatment of ischemia from arterial occlusion. The benefit of sympathectomy in patients with symptomatic Raynaud’s phenomenon is unclear, but it remains a potential intervention to assist healing of superficial ischemic ulcers and relieve rest pain in patients with Buerger’s disease.\(^\text{16}\)

### Hyperbaric Therapy

Scant objective evidence indicates that hyperbaric therapy alters the long-term course of chronic oblitative vascular disorders, presumably by accelerating formation of fine vessels. More success has been achieved with healing diabetic ischemic ulcers and salvaging ischemic skin grafts and flaps.\(^\text{17}\) Referral to a hyperbaric unit for chronic therapy should be made by the patient’s primary physician or vascular surgeon and not in the emergency department.

### SPECIFIC ARTERIOVASCULAR DISEASES

#### DISEASES OF CHRONIC ARTERIAL INSUFFICIENCY

**Arteriosclerosis Obliterans**

Arteriosclerosis obliterans (atherosclerotic occlusive disease, chronic occlusive arterial disease, oblitative arteriosclerosis) is the peripheral arterial presentation of atherosclerosis. Most often, arteriosclerosis obliterans affects the lower abdominal aorta, the iliac arteries, and the arteries supplying the lower extremities. Upper extremity manifestations are rare.

Arteriosclerosis obliterans is responsible for 95% of cases of chronic occlusive arterial disease. It is most common in persons older than 50 years, but as many as 19% of cases occur in patients aged 30 to 49 years. Men are affected more often than women (5:1 to 10:1). Approximately one third of patients with arteriosclerosis obliterans have coexistent coronary artery disease. The incidence of diabetes mellitus is 20 to 30%.\(^\text{18}\)

Risk factors for arteriosclerosis obliterans include cigarette smoking, hyperlipidemia, and hypertension. Of patients with arteriosclerosis obliterans, 70 to 90% are smokers when first examined, 75% have hyperlipidemia, and 30% have hypertension.\(^\text{18}\)

#### Clinical Features and Differential Diagnosis

Acute arterial occlusion from embolism, thrombosis, or trauma is ruled out primarily by history. Atheromatous emboli from proximal ulcerated plaques or aneurysms can cause small scattered ischemic lesions in the toes, feet, or legs, causing blue toe syndrome (Fig. 87-2). The peripheral pulses are present in blue toe syndrome. Exercise-induced claudication must be distinguished from the nocturnal muscle cramps that frequently occur during rest in elderly patients. Aortoiliac occlusive disease must be differentiated from osteoarthritis of the hip, which tends to be more variable from day to day, is not relieved completely with rest, and is not reliably reproduced by the same amount of exercise. Pseudoclaudication from the cauda equina syndrome is caused by narrowing of the lumbar canal from spondylosis, disease of the intervertebral disks, or spinal cord tumor. The symptoms mimic intermittent claudication but are less closely related to exercise and rest than true claudication.

The cause of lower extremity ulcers should be carefully determined. Approximately 5% of lower extremity ulcerations are caused by arterial insufficiency.\(^\text{19}\) These are usually located distal to the ankle, typically at the terminal portion of the digits, around the nail beds, or between the toes, caused by friction of one toe on another. Less common locations include the metatarsal heads, heel, and malleoli. Arterial insufficiency ulcers are painful but improve when the extremity is in a dependent position. They are associated with evidence of coexistent chronic arterial insufficiency (absence of hair growth on the dorsum of the feet, skin atrophy, absent pulses, and nail deformities). Ulcers are initially small, shallow, and dry. The base is gray, yellow, or black, with...
minimal or no granulation tissue. The rim of the ulcer is sharp and indolent, showing no signs of cellular proliferation or epithelialization.

Approximately 90% of lower extremity ulcers are caused by chronic venous insufficiency.19 These typically occur proximal to or in the region of the ankle, especially near the medial malleolus. Venous stasis ulcers are only mildly painful and improve with elevation of the extremity. Evidence of long-standing chronic venous insufficiency, including edema, prominent superficial veins, and stasis dermatitis, is present. Ulcers are moderate in size, with a weeping base and extensive granulation tissue. A rapidly developing ulcer is more suggestive of venous insufficiency.

Most of the remaining lower extremity ulcers are caused by diabetic neuropathy, alone or with arterial insufficiency.19 The location reflects sites of repeated trauma, including the toes, heels, and planter surface of the feet, especially the metatarsal heads. Neurotrophic ulcers are typically painless. Patients may have evidence of coexistent peripheral arterial insufficiency. The ulcers are deep and penetrating, often with suppurative drainage caused by an underlying infection or chronic osteomyelitis. Neurotrophic ulcers are usually surrounded by a rim of thick callus.

Hypertensive ulcers are rare and reflect long-standing, uncontrolled hypertension. These ulcers are typically near the lateral malleolus and start as painful, reddish blue areas of infarcted skin. A hemorrhagic bleb develops then breaks down into a superficial ulcer, which can reach a size of 5 to 10 cm. The ischemic ulcer has sharply demarcated borders, little granulation tissue, and minimal drainage. The pain is the most severe of all lower extremity ulcers.

Multiple ischemic ulcerations above and below the ankle suggest vasculitis or atheromatous embolization. Ulcers with regular edges in unusual locations may be factitial or may result from subcutaneous injection of illicit drugs. Thickened, rolled, and elevated edges with a central depression containing granulation tissue are characteristic of malignant ulcers.

Management

The first step is to identify patients whose symptoms are the sole result of arteriosclerosis obliterans without coexistent thromboembolic disease. Treatment for symptomatic patients depends on whether patients have functional ischemia or limb-threatening ischemia.20

Limb-threatening ischemia constitutes a surgical emergency. Angiography should be arranged to identify sufficiently localized disease to permit emergency bypass grafting.20 Patients with functional ischemia should have outpatient arrangements for noninvasive vascular testing or elective invasive contrast arteriography to determine treatment options such as bypass grafting. Ischemic ulcers or skin lesions should be cultured in the emergency department. Systemic antibiotics to cover skin organisms should be instituted if infection is present. Wet to dry dressings may help debride ulcers containing fibrin, debris, or infection. Radiographs of the underlying bones should rule out osteomyelitis. Patients with ischemic rest pain require hospitalization even if they are not surgical candidates. Bed rest, a warm environment, and maintenance of the limb in a dependent position usually relieve pain.

Buerger’s Disease (Thromboangiitis Obliterans)

First described by Buerger in 1908, thromboangiitis obliterans is an idiopathic inflammatory occlusive disease primarily involving the medium-sized and small arteries of the hands and feet.21 Patients are usually men aged 20 to 40 years who use tobacco, although recent reports indicate an increasing frequency of this disease in women. Buerger’s disease affects people of all races but is more prevalent in the Middle and Far East.22 The incidence in the United States is 20 per 100,000.23 The exact pathogenesis of Buerger’s disease is unknown, but virtually all patients are smokers.

Thromboangiitis obliterans is characterized by segmental acute and chronic inflammation in the smaller arteries of both upper and lower extremities. The initial arterial inflammatory process progresses to affect the adjacent veins and nerves, often leading to associated venous thrombosis and progressive fibrous encasement of these structures. There are painful, tender, reddened, or dark nodules over a peripheral artery with either a reduced or an absent pulse (phlebitis migrans).

Clinical Features

Clinical criteria for Buerger’s disease include (1) a history of smoking, (2) onset before the age of 50, (3) infraopopliteal arterial occlusive lesions, (4) either upper limb involvement or phlebitis migrans, and (5) absence of atherosclerotic risk factors other than smoking. A characteristic symptom of Buerger’s disease is foot or instep claudication caused by infraopopliteal arterial occlusion. Intense rubor of the affected extremity, particularly with dependency, is also characteristic. Foot pulses may be absent in the presence of normal femoral and popliteal pulses. Involvement of the hands is often bilateral and symmetrical, leading to the development of hand claudication or fingertip ulcers. Phlebitis migrans occurs early in the disease. Approximately 50% of patients experience Raynaud-type triphasic color response to cold. In the upper extremities, the digital arteries are usually more involved than the radial or ulnar arteries.22

Diagnostic Strategies

Adherence to diagnostic clinical criteria should be sufficient for emergency department diagnosis of Buerger’s disease. Noninvasive vascular laboratory testing can confirm the diagnosis and determine the extent of involvement. Although rarely required, angiography demonstrates multiple segmental occlusions.

Differential Diagnosis

Arteriosclerosis obliterans is most likely in patients older than 50 years who have signs of peripheral ischemia. In young women, autoimmune diseases, such as scleroderma or systemic lupus erythematosus, should be considered.22

Management

Permanent complete abstinence from tobacco is the only effective treatment for Buerger’s disease. If a patient does not completely stop smoking, alternating periods of quiescence are followed by exacerbations of severe arterial insufficiency. Patients who quit smoking have a benign clinical course. Despite this, many individuals who have Buerger’s disease continue to smoke, incurring severe pain at rest, tissue loss, and eventually amputation.

With early symptoms without threat of tissue loss, patient education and follow-up with a vascular surgeon are sufficient. Vascular surgery treatment options are varied for patients with severe symptoms or threatened tissue loss. Intractable pain can be controlled with epidural anesthesia. Intra-arterial or intravenous prostaglandin E1 and antithrombotic agents, including aspirin and heparin, have been used successfully.22 Patients with large-vessel arterial occlusion may benefit from arterial reconstruction. Symphactectomy is a potential treatment in advanced cases for cutaneous ulceration or relief of rest pain.24
Because patients with Buerger’s disease have good healing, intensive conservative treatment is usually successful in avoiding amputation.

### DISEASES OF ACUTE ARTERIAL OCCLUSION

#### Arterial Embolism

Despite advances in diagnosis and treatment, acute arterial embolus continues to cause substantial morbidity and mortality. Approximately 50% of acute arterial occlusions are caused by arterial embolism, and the incidence appears to be increasing. The other 50% are caused by in situ thrombosis.²

**Differential Diagnosis**

Phlegmasia cerulea dolens is a massive iliofemoral deep venous thrombosis. The initial symptom may be acute onset of a swollen and painful leg. As swelling continues, secondary arterial insufficiency with associated pallor (phlegmasia cerulea albans) may occur. In acute arterial embolism, leg swelling is usually absent, especially at the onset of pain. In addition, acute embolism produces a sharply demarcated pallor; phlegmasia cerulea dolens causes a cyanotic-appearing leg.

Aortic dissection may involve the arteries of the upper or lower extremity and may mimic acute embolus. A history of progressive severe pain, the presence of aortic insufficiency, and involvement at multiple sites suggest dissection. Acute neurologic syndromes (e.g., transverse myelitis, spinal subarachnoid hemorrhage, ruptured intervertebral disk) may produce sudden onset of unilateral or bilateral lower extremity weakness or sensory loss that mimics an acute aortic saddle occlusion.

Cold, blue extremities may result from low-output states such as hypovolemia, decreased cardiac output, dehydration, myocardial infarction, and pulmonary emboli in patients with long-standing atherosclerotic disease.

**Management**

Acute arterial embolism is a surgical emergency. The likelihood of limb salvage decreases after 4 to 6 hours. On the basis of clinical diagnosis alone, full doses of intravenous heparin should be administered immediately to minimize clot propagation. Patients whose history and physical examination findings clearly indicate an acute arterial embolism should undergo immediate Fogarty catheter embolectomy without prior angiography. In these patients, preoperative ultrasonography and angiography are rarely useful diagnostically and prolong the limb’s ischemic status.

If the differentiation of acute embolism and in situ thrombosis is uncertain, pretreatment angiography is usually diagnostic. Patients with acute emboli generally show minimal signs of atherosclerosis, occlusion at the site of an arterial bifurcation, sharply demarcated cutoffs, and lack of flow distal to the occlusion. In patients with in situ thrombosis, arteriography shows diffuse atherosclerosis, occlusion at sites other than arterial bifurcations, a tapered irregular cutoff, and well-developed collateral vessels. Emboli tend to lodge at arterial bifurcations, whereas arterial thrombi do not (see Table 87-1).

Intra-arterial thrombolytic therapy for acute embolism remains investigational. Immediate limb-threatening ischemia precludes consideration of treatment with thrombolytic therapy in most patients. Potential risks of thrombolytic therapy in arterial embolism patients with non–limb-threatening ischemia include partial clot lysis with distal embolization or recurrent embolic events from the primary source of the initial embolus.²³

#### Atheroembolism (Blue Toe Syndrome)

Atheroemboli are microemboli consisting of cholesterol, calcium, platelet aggregates, and hemorrhagic debris that break off from proximal atherosclerotic plaques or aneurysms and lodge in distal end arteries. In the central nervous system, atheroembolism causes transient ischemic attacks and strokes. In the peripheral vascular system, atheroemboli characteristically are found in the lower extremities with cool, painful cyanotic toes in the presence of palpable distal pulses (see Fig. 87-2).

**Clinical Features**

The typical presentation of atheroembolism is the sudden onset of a small painful (cyanotic and tender) area on the foot, typically the toe.²⁴ If bilateral involvement is present, the distribution is not symmetrical. Posterior tibial and dorsalis pedis pulses are present. The physical examination should be directed toward identification of a proximal source, such as an atherosclerotic aneurysm in the aorta or iliac, femoral, or popliteal artery.

**Differential Diagnosis**

A variety of conditions can mimic the blue toe syndrome. Acrocyanosis is painless, has a symmetrical distribution, and is located in the hands, nose, and lips. Poor peripheral perfusion from low cardiac output must also be considered. Vasculitis typically causes palpable purpuric lesions and is associated with constitutional symptoms of low-grade fever, myalgias, and weight loss. Previous frostbite may leave the extremities sensitive to cold. Local injury to the foot of the diabetic patient is easily differentiated.

**Management**

Treatment is directed toward identifying and removing the proximal source of atheroembolism. Angiography is the most accurate diagnostic method for determining the source of emboli. If the source is an aortic aneurysm and the patient is a surgical candidate, operative repair should be performed. Stenotic lesions in the iliac or femoral arteries can be treated with local endarterectomy, vascular bypass, or angioplasty.¹⁴ Medical management with aspirin, dipyridamole, crystalline warfarin sodium (Coumadin), or steroids has variable results.

#### Arterial Thrombosis

Approximately 50% of acute arterial occlusions are caused by in situ thrombosis.² Acute arterial thrombosis is almost always superimposed on a complicated atherosclerotic lesion but can be caused by vasculitis or trauma. With limb-threatening ischemia, angiography can be used to evaluate the feasibility of emergency bypass grafting. In patients with non–limb-threatening ischemia, angiography may be required to distinguish acute embolism from thrombosis (see Table 87-1).

**Management**

Heparinization should be started when the diagnosis is made. Patients with severe limb-threatening ischemia require emergency direct or Fogarty catheter thrombectomy combined with bypass grafting. Simple thrombectomy alone often fails as a result of rethrombosis. Patients who have atherosclerotic disease not amenable to vascular bypass, who are too ill to tolerate revascularization, or who have irreversible ischemia require primary amputation. Patients with non–limb-threatening ischemia are best treated nonoperatively with heparin and low-dosage intra-arterial thrombolytic therapy.
PERIPHERAL ARTERIAL ANEURYSMS

A true aneurysm is an abnormal localized dilation of the intact wall of any vessel caused by a combination of mural weakness and hemodynamic forces. Aneurysms enlarge at a rate determined by the cause. Those caused by atherosclerosis progress slowly over years; those caused by trauma or infection enlarge over days, weeks, or months. The primary risk of central aneurysms (abdominal aorta, iliac arteries, and visceral arteries) is rupture (see Chapter 84). Peripheral arterial aneurysms rarely rupture; instead, they are complicated by thrombosis or embolism that jeopardizes distal tissues.25

The cause of an aneurysm depends on its anatomic location. Lower extremity aneurysms are most often atherosclerotic in origin. Upper extremity aneurysms are usually caused by local trauma. Visceral aneurysms are from abnormal hemodynamics, atherosclerosis, or infectious causes.

Lower Extremity

Femoral and popliteal artery aneurysms almost always occur in older men with advanced atherosclerosis. Twenty-five percent of patients have distal atheroembolism or thromboembolism; an additional 15% have total occlusion from in situ thrombosis.25

Popliteal aneurysms are the most common peripheral aneurysms and occur bilaterally in approximately 60% of patients.25 An abdominal aortic aneurysm occurs in almost 80% of patients with bilateral popliteal aneurysms. Most patients have claudication, thromboembolic events, atheroembolic events, or gangrene. Aneurysmal dilation can cause venous compression with associated deep venous thrombosis.

Femoral aneurysms are the second most common peripheral aneurysms and manifest similarly to popliteal aneurysms. Femoral aneurysm dilation can also compress the femoral nerve, producing anterior thigh pain or weakness.

Diagnosis of both popliteal and femoral aneurysms is by palpation of a pulsatile mass. Plain radiographs may show unilateral or bilateral calcified aneurysms. Ultrasonography and CT are diagnostic. Arteriography yields definitive diagnosis and indicates involvement of distal vessels. Patients with a lower extremity aneurysm should be evaluated for the presence of other aneurysms.

Asymptomatic patients can undergo elective surgical excision of the aneurysm and end-to-end anastomosis or graft interposition. Simultaneous repair of coexisting abdominal aorta or contralateral extremity aneurysms combined with vascular bypass is typically done. Patients with limb-threatening thromboembolic events are first treated with Fogarty catheter embolectomy.25

Upper Extremity

Atherosclerosis generally spares the upper extremities, so peripheral arterial aneurysms in the upper extremities are rare, making localized trauma the most common cause.

The causes of proximal subclavian artery aneurysms are thoracic outlet obstruction, trauma, and, rarely, atherosclerosis. Subclavian aneurysms from atherosclerosis represent severe disease, and 30 to 50% of patients so afflicted also have aortoiliac or other peripheral aneurysms.26 Symptoms depend on the aneurysm’s anatomic location. Patients may have chest, neck, or shoulder pain from acute expansion. Compression of the right recurrent laryngeal nerve can lead to voice change. Compression of the trachea can lead to stridor or other respiratory complaints. The chest radiograph may reveal a superior mediastinal mass, easily confused with a neoplasm.

The subclavian artery can be compressed by a complete cervical rib that articulates with the first rib, producing a poststenotic dilation in the proximal subclavian and distal axillary artery. This syndrome occurs more often in women and in the dominant upper extremity. Cervical ribs occur in 0.6% of the population.27

Axillary artery aneurysms are most often caused by blunt trauma from inappropriate and prolonged use of crutches. Humerus fracture and anterior shoulder dislocation are less common causes.26

Subclavian, subclavian-axillary, and axillary artery aneurysms share the common complications of thromboembolism and limb-threatening ischemia, neuromuscular and sensory dysfunction from brachial plexus compression, and central nervous system ischemia produced by retrograde thromboembolism in the vertebral and right carotid circulation. A systolic bruit with a palpable thrill is common.

Arteriography to confirm the diagnosis and determine involvement of distal vessels is the diagnostic procedure of choice. Surgical treatment consists of aneurysm resection, vascular grafting, and reestablishment of arterial continuity.

The rare syndrome of ulnar artery aneurysm (hypothenar hammer syndrome) is associated with occupational trauma in which the heel of the palm is used to hammer, push, or twist objects.28 Patients are often mechanics, carpenters, and machinists.

The ulnar artery fits snugly into the bony canal at the hypothenar eminence under the hook of the hamate bone. Long-term repetitive damage to this region results in aneurysm formation.28 The aneurysm may develop a mural thrombus that repeatedly embolizes to the superficial palmar arch or to a digital artery. Symptoms consist of paresthesias, pain, coolness, and cyanosis, most often in the little and ring fingers and occasionally in the middle and index fingers. The thumb is characteristically spared because of its radial artery blood supply. Diagnosis is easily made by finding a pulsatile or nonpulsatile tender mass in the hypothenar eminence of the dominant hand. The Allen test may demonstrate occlusion of the ulnar artery. Angiography of the distal vessels is diagnostic. Proximal angiography rules out the subclavian and axillary arteries as embolic sources. Treatment requires surgical resection of the aneurysm and reestablishment of ulnar artery continuity. Adjunctive preoperative fibrinolytic therapy may be helpful.28

Viscera

Splenic Artery Aneurysms

Splenic artery aneurysms account for 60% of all visceral arterial aneurysms. They are the only aneurysms that are more common in women, with a female-to-male ratio of 4:1.27 The development of aneurysms in the splenic artery has been attributed to systemic arterial fibrodysplasia, portal hypertension, and increased splenic arteriovenous shunting that occurs in pregnancy.

Splenic artery aneurysms are most often asymptomatic. Symptomatic patients exhibit vague left upper quadrant or epigastric discomfort and occasional radiation of pain to the left shoulder or subscapular area. Because most splenic artery aneurysms are less than 2 cm in diameter, a pulsatile mass is not palpable. Occasionally, a systolic bruit can be heard.

Only 2% of splenic artery aneurysms result in life-threatening rupture.29 More than 95% of ruptures occur in young women during pregnancy and can be confused with ectopic pregnancy or placental abruption.

Splenic artery aneurysms are usually an incidental discovery on the abdominal radiograph as signet ring calcifications in the left upper quadrant. Ultrasonography, CT, and MRI can distinguish aneurysms from other cystic lesions in the left upper quadrant.29 An angiogram is usually required to confirm the diagnosis. Symptomatic splenic artery aneurysms require immediate operative
intervention, particularly in pregnant women or in women of childbearing age. The rate of maternal mortality from rupture during pregnancy is approximately 70%. In asymptomatic patients, transcatheter embolization is an alternative to surgery.30

Hepatic Artery Aneurysms

Hepatic artery aneurysms represent 20% of visceral artery aneurysms. The lesions are caused by atherosclerosis, infection (most often as a complication of intravenous drug abuse), major abdominal trauma, and polyarteritis nodosa. Hepatic artery aneurysms affect men twice as often as women and usually occur after 60 years of age.

Most aneurysms remain asymptomatic, but unruptured symptomatic aneurysms generally produce symptoms consistent with cholecystitis: vague, persistent, right upper quadrant or epigastric pain radiating to the back. Large aneurysms can cause severe upper abdominal discomfort, similar to pancreatitis. Hepatic artery aneurysms may rupture into the common bile duct, peritoneum, or adjacent hollow viscera, with a mortality rate of 35%.

An abdominal bruit or palpable pulsatile mass is usually not present on physical examination. Aneurysmal calcification may be seen on a plain abdominal radiograph, but the diagnosis can be made reliably by angiography. Ultrasonography and CT can detect asymptomatic hepatic artery aneurysms.29

Because of the high mortality rate with aneurysmal rupture, an aggressive approach to management is warranted. Surgical resection of the aneurysm is performed in operative candidates. Transcatheter occlusion can be used in patients who are high surgical risks.31

Superior Mesenteric Artery Aneurysms

Superior mesenteric artery aneurysms are the third most common visceral aneurysms. Nearly 60% are infected aneurysms caused by nonhemolytic streptococci from left-sided bacterial endocarditis. Atherosclerosis and trauma are much less common causes. Patients are usually younger than 50 years of age; men and women are affected equally.

Patients generally have intermittent upper abdominal pain consistent with abdominal angina. Fifty percent have a pulsatile abdominal mass on physical examination. The stigmata of subacute bacterial endocarditis may be present. Plain abdominal radiographs may show a calcified aneurysm. Angiography is necessary to confirm the diagnosis.

Management of superior mesenteric artery aneurysm should address any underlying infectious process. The surgical approach is difficult and varies with the condition of the patient, the shape of the aneurysm (saccular or fusiform), and the intraoperative assessment of bowel viability.

Infected Aneurysms

Mycotic Aneurysms

The term mycotic aneurysm is a source of confusion because there is no association with fungal disease. Although the term has been used to describe any infected aneurysm regardless of cause, it should be reserved for infected aneurysms resulting from bacterial endocarditis, as originally described in 1885 by Osler.32,33

Septic emboli from infective endocarditis implant in one of two ways. First, hematogenous seeding of bacteria can occur in nonaneurysmal arteries whose vessel walls have been damaged by preexisting atherosclerosis. Second, septic emboli can become lodged in the vasa vasorum of larger vessels, causing vessel wall ischemia and infection. In smaller vessels, septic emboli tend to lodge at arterial bifurcations, arteriovenous fistulae, or sites of arterial stenosis. Mycotic aneurysms are most common in the aorta, superior mesenteric artery, and intracranial and femoral arteries.

The infecting organism in mycotic aneurysms reflects the bacteriology of infective endocarditis. Viridans streptococci are the most common organisms, although intravenous drug abusers are most often infected by Staphylococcus aureus. Patients who have mycotic aneurysms tend to be 30 to 50 years of age. The mortality rate is 25% (Table 87-2).32,33

Atherosclerotic Arteries

Currently, the most common cause of an infected aneurysm is sepsis with hematogenous spread of bacteria, such as Salmonella, Staphylococcus, and Escherichia coli, to atherosclerotic arteries. Large vessels (especially the aorta) rather than peripheral arteries are the most common site. Patients tend to be older than 50 and to have well-established atherosclerosis. Perforation often occurs before diagnosis and carries a mortality rate of 75%.33

Preexisting Aneurysms

The incidence of infection in patients with preexisting atherosclerotic aneurysms is estimated at 3 to 4%, and patients with

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<th>Table 87-2 Clinical Characteristics of Infected Aneurysms</th>
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<td>MYCOTIC ANEURYSM</td>
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ruptured aneurysms have a higher incidence of positive bacterial culture results than those who have elective surgical treatment of an asymptomatic aneurysm. Gram-positive organisms, especially Staphylococcus, predominate (60%). The rate of mortality is extremely high (90%) because of aneurysm rupture.32,33

**Post-traumatic Pseudoaneurysms**

Post-traumatic infected aneurysms result from invasive hemodynamic monitoring, angiography, and intravenous drug use. The most common artery affected is the femoral because of its involvement in groin injection. S. aureus is isolated in 30 to 70% of cases. Because of the more peripheral location and early identification, the mortality rate is low (5%).34

The clinical presentation of an infected aneurysm varies with anatomic location and underlying pathophysiologic process. Patients with infected abdominal aneurysms are often misdiagnosed. Onset is usually insidious; low-grade fever may be present for several months. Common findings are fever (75%), back and abdominal pain (33%), and palpable aneurysm (53%). More peripheral aneurysms, especially infected femoral pseudoaneurysms, are characterized by a tender groin mass, some manifestation of sepsis, or bleeding.34 Almost all are easily palpable. Although rare, fungal infections should be considered in patients who are chronically immunosuppressed, have been treated recently for disseminated fungal disease, or have diabetes mellitus.35,36

Positive blood cultures in a patient with a preexisting aneurysm should prompt treatment as an infected aneurysm until disproven. Bacteremia often is continuous, and blood cultures are positive in approximately 70% of cases, so negative blood cultures do not rule out this diagnosis. Angiography should be performed when an infected aneurysm is suggested.35 Indium-111–labeled white blood cells can confirm or rule out infected aneurysms.35

Treatment includes both antibiotics and surgical repair. Antibiotic therapy is usually continued for at least 6 to 8 weeks, although some physicians advocate lifelong treatment after successful surgical repair.35 The most important intervention is timely repair.35,33 Without surgery, aneurysm rupture with exsanguinating hemorrhage is inevitable.33

**Traumatic Aneurysms**

Traumatic aneurysm refers to a pseudoaneurysm that follows perforation of the arterial wall, with formation of a perivascular hematoma. Chronic traumatic aneurysms may or may not be associated with an arteriovenous fistula. Pseudoaneurysm is a synonym for false aneurysm.

The usual presentation is a pulsatile mass found near the course of an extremity artery, with a history of trauma more than 1 month earlier.39 The expanding aneurysm may compress associated peripheral nerves and produce neuropathy. Distal perfusion is usually well maintained, and thromboembolism is rare. A loud systolic and possibly a separate faint diastolic murmur are characteristic.

The diagnosis can be verified with conventional angiography, digital subtraction arteriography, or CT. Surgical excision of the aneurysm is indicated as soon as possible to decrease the risk of complications, including rupture, thrombosis, or neurologic dysfunction caused by continued expansion.

**VASOSPASTIC DISORDERS**

Vasospastic disorders are characterized by an abnormal vasomotor response in the distal small arteries. Blood flow in the peripheral circulation is controlled by local, autonomic, and humoral mechanisms.9 The cause of the increased vasospastic response is unknown.

Raynaud’s disease is the most common vasospastic disorder and occurs five times more often in women than in men. By definition, cases of Raynaud’s disease have no evidence of an underlying cause. The diagnosis is correct in 95% of cases that meet these criteria: (1) episodes are precipitated by cold or emotion; (2) symptoms are bilateral; (3) gangrene is absent or is minimal and confined to the skin; (4) no disease or condition that could cause a secondary Raynaud’s phenomenon is present; and (5) symptoms have been occurring for at least 2 years.40

The classic Raynaud’s attack is triphasic: the fingers become white, then blue, and finally red. This is produced initially by complete closure of the palmar and digital arteries (and possibly arterioles), producing cessation of capillary perfusion. When a slight relaxation of arterial spasm occurs, a slight flow of blood returns into the dilated capillary bed, where it rapidly dissipates, producing cyanosis. Arterial spasm usually spontaneously resolves, arterial flow returns to baseline, but reactive hyperemia produces a red extremity. Attacks are often precipitated by cold and emotional stress. Raynaud’s disease usually has a benign course. True histologic changes within the vessel wall are absent. Reassurance, education, and primary care follow-up are the only treatment necessary for true Raynaud’s disease.

Raynaud’s phenomenon is the term given to Raynaud’s disease when there is an identifiable underlying causative disorder. Connective tissue disorders, including scleroderma, rheumatoid arthritis, and systemic lupus erythematosus, have the highest association with Raynaud’s phenomenon. Treatment should be directed toward identifying the underlying disorder and minimizing threatened tissue loss if present.40

Benign livedo reticularis is caused by spasm of the dermal arterioles and may involve all parts of the upper and lower extremities, and the trunk. It is most common when skin is exposed to a cool environment. It is never associated with histologic vascular abnormality and quickly resolves when the exposed skin is covered or the environment is warmed. Other conditions, similar to the causes of Raynaud’s phenomenon, can have secondary livedo reticularis along with other peripheral vascular disease manifestations.40

Acrocyanosis is the least common of the vasospastic disorders and is characterized by persistent, painless, symmetrical cyanosis of the fingers, the hands, and less often the feet. The disease is benign and not associated with either vascular abnormality or an underlying disorder. Pain, trophic skin changes, and ulceration do not occur. This disorder occurs more often in women, is intensified by exposure to cold, and decreases with warming. The diagnosis is made by the bilateral and persistent nature of the findings, localized to the hands or feet in the presence of normal arterial pulses. The involved extremities are nearly always cold, and excessive perspiration is common. Except for reassurance and protection from cold, treatment is usually unnecessary.40

Primary erythromelalgia is a rare syndrome of paroxysmal vasodilation with burning pain, increased skin temperature, and redness of the feet and less often the hands. However, secondary erythromelalgia can occur in patients with underlying disease processes, most often systemic lupus erythematosus, myeloproliferative disorders, hypertension, venous insufficiency, or diabetes mellitus. Erythromelalgia is as common in children as adults, but in children it is less likely to be associated with an underlying systemic illness. Attacks are not triggered by cold and usually occur in modest ambient temperatures. Skin temperature of the involved digits is high compared with the patient’s core temperature. Symptoms may remain mild for years or may become disabling. Tissue loss and trophic skin changes do not occur. Although rest, elevation of the extremities, and cold compresses or immersion in ice can provide temporary relief, no consistently effective treatment has been found for the multiple, often daily episodes of pain that occur with erythromelalgia.40
Thoracic outlet syndrome involves compression of the brachial plexus, subclavian vein, or subclavian artery at the superior aperture of the thorax. Thoracic outlet syndromes were previously categorized by cause as scalenus anticus, costoclavicular, hyperabduction, cervical rib, and first thoracic rib syndromes. They are now most easily divided into three types—neurologic, venous, and arterial—depending on the predominant symptoms.

Compression of the brachial plexus causes the neurologic type of thoracic outlet syndrome and accounts for approximately 95% of all cases. Symptoms begin between the ages of 20 and 50 years, with women predominating at a ratio of about 3:1. Compression or thrombosis of the subclavian vein constitutes the venous type of thoracic outlet syndrome and is responsible for 4% of all cases. It occurs most often in men 20 to 35 years of age. The arterial type of thoracic outlet syndrome is rare, occurring in approximately 1% of all cases, but is potentially the most serious of the three types. Men and women are equally affected in a bimodal age distribution of young adults (from cervical rib compression) and patients older than age 50 (from localized atherosclerosis caused by arterial compression). Figure 87-3 demonstrates the relationship between anatomic abnormalities and neurovascular compression.

Principles of Disease

Roos has described four basic concepts of thoracic outlet syndromes: (1) patients who have a thoracic outlet syndrome develop an anatomic abnormality predisposing them to symptoms under certain conditions; (2) brachial plexus compression or irritation constitutes approximately 95% of all thoracic outlet syndrome cases and is rarely caused by compression of the subclavian artery; (3) bedside testing for thoracic outlet syndrome based on positional compression of the subclavian artery is insensitive and unreliable; and (4) in advanced or refractory cases, the causative anatomic abnormalities must be surgically corrected.

The subclavian artery courses over the first rib between the scalenus anticus muscle anteriorly and the scalenus medius muscle posteriorly, when passing under the clavicle to the axilla, where the brachial plexus lies posteriorly and laterally. Four anatomic abnormalities have been associated with thoracic outlet syndrome.

Cervical rib syndrome results from an uncommon abnormality (0.5-0.7% of all chest radiographs), which is bilateral in 70% of patients. It occurs twice as often in women as men. Most cervical ribs are incomplete, attached to a fibrous band on the scalene tubercle of the first rib. The site of compression is the scalene hiatus, made up of the scalene anterior muscle anteriorly, the scalene medius posteriorly, and the cervical rib inferiorly.

Scalenus anticus syndrome results when the neurovascular bundle is compressed by various insertions of the anterior scalene muscle as it passes through the interscalene triangle. In some patients the subclavian artery passes through the body of the muscle.

Costoclavicular syndrome results when the shoulders are moved backward and downward. Causes include hypertrophy of the
subclavius muscle, abnormalities of the first rib, and past clavicular fractures.

Hyperabduction syndrome results from the neurovascular compression that occurs when the arms are placed in the hyperabducted position. The site of compression is in the retroclavicular space anterior to the first rib or at the point where the neurovascular bundle passes beneath the pectoralis minor muscle.

The neurologic and venous compression type of thoracic outlet syndrome can be associated with any underlying anatomic abnormality. Bony abnormalities (cervical rib, first thoracic rib, or clavicle) are the most common causes of the arterial type of thoracic outlet syndrome (Fig. 87-4A).

Clinical Features

Compression or irritation of the brachial plexus most often affects the lower two nerve roots, eighth cervical (C8) and first thoracic (T1), producing pain and paresthesias in the ulnar nerve distribution. The second most common anatomic pattern is involvement of the upper three nerve roots of the brachial plexus (C5, C6, and C7), with symptoms referable to the neck, ear, upper chest, upper back, and outer arm in the radial nerve distribution. Venous compression eventually progresses to intimal damage and subclavian vein thrombosis, with venous engorgement and swelling of the affected extremity. Persistent subclavian artery compression eventually results in poststenotic aneurysm formation and its sequelae.

Physical Examination

The Adson, costoclavicular, and hyperabduction maneuvers are unreliable as diagnostic tests.42 The most reliable test in screening for thoracic outlet syndrome is the elevated arm stress test (EAST).43 With the patient sitting, the arms are abducted 90 degrees from the thorax and the elbows flexed 90 degrees, with the shoulders braced slightly behind the frontal plane. The patient is asked to open and close the fists slowly but steadily for a full 3 minutes and to describe any symptoms that develop. Normal patients perform this test without symptoms other than mild fatigue. The patient with thoracic outlet syndrome, however, usually has early heaviness and fatigue of the involved limb, gradual onset of numbness of the hand, and progressive aching through the arm and top of the shoulder. Within the 3 minutes the patient usually drops the hand to the lap for relief of the progressive, crescendo distress that becomes intolerable. Patients with carpal tunnel syndrome may experience dysesthesias in the fingers but do not have shoulder or arm pain. Patients with cervical disk syndromes may have pain in the neck and shoulder but have no arm or hand symptoms.

The EAST evaluates all three types of thoracic outlet syndrome: neurologic, venous, and arterial. Radial pulses can be palpated by the examiner during the test. The presence of a radial pulse and a positive EAST test result are strong indications that the basis of symptoms is neurologic involvement of the brachial plexus.

The hands should be observed for changes in skin color, warmth, moisture, or muscular atrophy. Triceps muscle strength (innervated by C7) should be tested bilaterally. Muscle strength of the interosseous muscles (innervated by C8 and T1) should be tested by asking the patient to spread the fingers apart against resistance. The muscles innervated by the radial nerve are tested by having the patient hyperextend the thumb and dorsiflex the wrist against resistance. The median nerve innervates the thenar muscles, which can be tested by asking the patient to abduct the thumb away from the palm with the thumb pointing straight to the ceiling. Tinel’s sign (“electric shock” to tips of fingers) is an indication of carpal tunnel compression of the median nerve and is elicited by percussing the volar aspect of the wrist. Gentle pressure with the thumb in the supraclavicular fossa over the brachial plexus may reproduce thoracic outlet symptoms after several seconds. The cervical spine and upper extremity reflexes should be assessed.

A blood pressure difference between the two arms is a reliable indication of arterial involvement. The blood pressure in the affected arm is lower. Doppler ultrasonography may be helpful in demonstrating comparatively reduced pressure over the pairs of radial, ulnar, and brachial arteries. The supraclavicular area should be auscultated bilaterally for subclavian bruits.

Ancillary Evaluation

Cervical spine radiographs with oblique views and chest radiographs are used to identify skeletal abnormalities (first rib, cervical rib, clavicle deformity), trauma, arthritis, spondylitis, Pancoast tumor, or other pulmonary disease. Electromyography, nerve conduction times, and somatosensory evoked potentials are generally unreliable and do not provide objective evidence of thoracic outlet syndrome.42 Patients thought to have cervical disk or spinal cord disease may require cervical myelography, CT, or MRI.

Arteriography is recommended with (1) obliteration of radial pulse on the EAST, (2) blood pressure 20 mm Hg less than that of the opposite asymptomatic limb, (3) possible subclavian stenosis or aneurysm (bruit or abnormal suprascapular pulsation), and (4) evidence of peripheral emboli in the upper extremity.42 Venography is indicated if the patient has a history of intermittent or persistent edema of the hand or arm, peripheral unilateral cyanosis, or a prominent venous pattern over the arm, shoulder, or chest.43

Differential Diagnosis

The differential diagnosis of thoracic outlet syndrome includes herniated cervical disk, cervical spondylitis, spinal cord tumor, ulnar nerve compression at the elbow, carpal tunnel syndrome, orthopedic shoulder problems (sprain, rotator cuff injury, tendinitis), trauma, postural palsy, angina pectoris, and a variety of neuropathies, including those associated with multiple sclerosis, alcoholism, and diabetes.

Patients with a herniated cervical disk have more severe persistent pain radiating in a sharply demarcated dermatomal distribution (usually C4-5 or C5-6) and often have localized tenderness of the cervical spine at the affected level. Carpal tunnel syndrome is characterized by nocturnal symptoms of pain and paresthesias and an associated Tinel’s sign on physical examination. Brachial plexus compression and irritation can be confused with other vascular conditions, such as Raynaud’s disease, vasospastic disorders, vasculitis, or arterial ischemia. Unilateral symptoms should suggest thoracic outlet syndrome, whereas bilateral symptoms suggest a systemic process. Subclavian or axillary venous thrombosis from thoracic outlet syndrome must be differentiated from...
thrombophlebitis or mediastinal venous obstruction from a benign or malignant process (Pancoast tumor).

Management

Treatment varies depending on whether the involvement is neurologic, arterial, or venous. In patients with only brachial plexus involvement and with minimal signs and symptoms, conservative treatment with physiotherapy and shoulder girdle exercises is sufficient. Surgery is reserved for patients with intolerable pain or progressive loss of function and strength of the arm or hand. First rib and anomalous muscle or fibrous tissue resection provides consistent relief of symptoms and minimal morbidity (see Fig. 87-4B).

Patients with arterial complications of thoracic outlet syndrome (thrombosis, thromboembolism, or acute ischemia) require immediate heparinization and angiography; Fogarty catheter embolectomy, if appropriate; and emergency or urgent surgical exploration. Patients with axillary and subclavian vein thromboses require immediate heparinization and venography and are treated with surgical thrombectomy or systemic fibrinolytic therapy.

Disposition

The correct diagnosis of thoracic outlet syndrome can be achieved in more than 90% of patients with a careful history, physical examination, and bedside testing alone. Neurologic, orthopedic, or vascular surgery consultation is indicated according to the pathologic condition.

PERIPHERAL ARTERIOVENOUS FISTULAE

Acquired peripheral arteriovenous fistulae are most often caused by trauma (gunshot wounds, stab wounds, or surgery), with malignancy, infection, and arterial aneurysms as less common causes. Patients generally seek medical care several months after an invasive surgical procedure or penetrating injury.

Differential Diagnosis

The correct diagnosis of an arteriovenous fistula can usually be made with clinical examination alone. A constant systolic and diastolic (to-and-fro) murmur associated with a palpable thrill is characteristic. Sixty percent of arteriovenous fistulae are also associated with a coexisting false aneurysm. Patients with peripheral venous disease may have similar cutaneous manifestations (varicose veins and stasis pigmentation) but lack vascular bruits. Infection in the form of bacterial endarteritis may complicate large fistulae.

Management

Acquired peripheral arteriovenous fistulae usually increase in size with time if surgery is delayed. Vessel dilation, peripheral ischemia, and cardiac output increase. Transthoracic embolization with detachable balloons and liquid acrylic tissue adhesives (e.g., isobutyl 2-cyanoacrylate) is used for surgically inaccessible fistulae.

VASCULAR ABNORMALITY CAUSED BY DRUG ABUSE

Principles of Disease

The vascular complications of parenteral drug use have risen significantly in both frequency and severity since the late 1980s. These intravenous or intra-arterial injuries can result in acute arterial ischemia, infected pseudoaneurysms, lymphatic obstruction, or direct neurologic injury.

Acute arterial ischemia results from direct drug effects or endogenous catecholamine release after injection. Endothelial wall damage can stimulate platelet aggregation and thrombus formation. Precipitated crystals, talc, or foreign body emboli can cause arterial occlusion. Necrotizing arteritis can produce ischemia and is especially prevalent in patients who abuse intravenous methamphetamine.

Infected pseudoaneurysms associated with arteriovenous fistulae result from a through-and-through puncture of the artery with simultaneous contamination from either skin flora or contaminated needles or drug. These fistulae are the most common vascular lesions resulting from intravenous drug abuse. Secondary infection of the vascular structure may be covered by surrounding soft tissue infection (cellulitis or abscess). Infected aneurysms at sites distant from the injection can occur.

Intravenous drug abusers can develop unilateral hand edema or “puffy hand syndrome” because of gradual obliteration of the superficial venous vessels and chronic lymphatic obstruction. Direct injury to adjacent nerves, polyneuritis, and ischemic neuritis can result from intravenous drug abuse. Coexisting infections include cellulitis, septicemia, and bacterial endocarditis.

Clinical Features

Patients may withhold information about the use of intravenous drugs, but objective evidence such as track marks may be present. Distal ischemia after intra-arterial injection most often occurs in the upper extremity after injection of the brachial or radial artery. The immediate onset of a severe, burning pain at the time of injection is a characteristic hallmark. Patients have a painful, edematous upper extremity with patchy blue-purple skin discoloration. Distal pulses are generally present, but the skin temperature of the involved extremity is decreased. Because patients tend to seek attention early, the site of injection may be identifiable over the radial or brachial artery. Evidence of gangrene, pregangrenous changes, or neuromuscular deficits may accompany this syndrome.

Patients with infected pseudoaneurysms have a painful mass develop several days to weeks after injection, with resultant bleeding or “hitting pink.” The mass is usually pulsatile, and 50% have an associated bruit. Infected pseudoaneurysm is part of the differential diagnosis of cutaneous abscess or cellulitis in an intravenous drug user. Infected pseudoaneurysms are most often encountered in the lower extremities (80%). All patients should be carefully evaluated for sepsis, metastatic infection, and bacterial endocarditis. A peripheral vascular examination with careful documentation of pulses should be performed. A radiograph of the affected extremity can detect subcutaneous needle fragments or foreign body. Angiography is the diagnostic procedure of choice for suggested pseudoaneurysm or distal ischemia. Ultrasonography is often unable to distinguish an aneurysm from an abscess or cellulitis.

Management

Therapeutic considerations for acute ischemia from intra-arterial injection are primarily conservative. Intra-arterial vasodilators, heparin, low-molecular-weight dextran, fibrinolytic therapy, analgesics, systemic warming to stimulate vasodilation, antibiotics, elevation of the affected limb to promote venous drainage, and physical therapy have not significantly altered the outcome or amputation rate in this patient population. Surgical treatment is reserved for delayed amputation, with the goal of preserving as much tissue as possible. Gradual resolution of
symptoms without surgical intervention is the most common outcome.

Patients with infected pseudoaneurysms require aneurysm resection, debridement of infected tissue, and ligation of the proximal and distal uninfected arteries. Autogenous vein bypass through uninfected tissue planes may require an extensive surgical approach. Intravenous nafcillin is recommended for mild infections, nafcillin and a second- or third-generation cephalosporin for major infections, and vancomycin and a second- or third-generation cephalosporin or an aminoglycoside for patients who are bacteremic or overtly septic. Methicillin-resistant *S. aureus* and gram-negative rods are increasing in frequency as the causative agents in infections and vascular injury resulting from drug abuse, and vancomycin should be added if these organisms are suspected.

**PROBLEMS RELATED TO LONG-TERM CENTRAL VENOUS ACCESS**

**Hickman-Broviac Catheter**

The Hickman-Broviac double-lumen catheter is in common use, with the smaller Broviac line used for the administration of intravenous therapy and the larger Hickman line reserved for additional venous access and blood withdrawal (Fig. 87-5; see also Fig. 87-7B). This catheter is generally inserted into the cephalic, subclavian, external, or internal jugular vein, with the distal tip just above the right atrium. The proximal end exits through a subcutaneous tunnel from the lower anterior chest wall. A felt cuff (Dacron) is used to anchor it in place subcutaneously. The Hickman-Broviac catheter is made of polymeric silicone rubber that is of low thrombogenic potential but extremely flexible and soft. Because of the pliability of the material, the catheter must be treated gently. Clearing an obstructed catheter with a guidewire is increasing in frequency as the causative agents in infections and vascular injury resulting from drug abuse, and vancomycin should be added if these organisms are suspected.

**Routine Care and Use**

The smaller Broviac line is most often used for the infusion of total parenteral nutrition or fat emulsions. This line should be irrigated with 6 mL of normal saline solution between different infusions to prevent mixing of incompatible solutions, development of precipitation, and resultant catheter occlusion. The larger Hickman line should be used to withdraw blood. This line should be irrigated with 6 mL of heparinized saline after blood withdrawal to prevent clot formation in the catheter lumen. When a clamp is used, it should be placed over a piece of tape wrapped around the line. The clamp should have a smooth surface, because teeth or prongs could sever or abrade the line.

Routine care and frequency of catheter dressing changes vary with the preference of the treating physician. Most patients become skilled in routine catheter maintenance and are a reliable source of information. Absolute sterile technique is essential when the catheter is manipulated.

**Catheter Occlusion**

Hickman catheters can exhibit complete or partial obstruction to flow in either line. Complete obstruction, in decreasing order of frequency, results from (1) clots within the catheter lumen, (2) precipitates within the catheter lumen, and (3) mechanical obstruction. In catheters that accept infusions at normal rates but cannot be aspirated, the causes, in decreasing order of frequency, are (1) catheter lodged against the wall of the vessel, (2) occluding fibrin sheath around the catheter tip, (3) ball valve or mural thrombus, and (4) central venous thrombosis. Patients who have intermittent complete occlusion and withdrawal occlusion have a type of mechanical obstruction called *pinch-off syndrome*, in which the catheter lumen is compromised from mechanical forces acting on it between the clavicle and the first rib. Clots within the catheter lumen, obstructing fibrin sheaths, and ball valve or mural thrombus often respond to low-dose intracatheter urokinase, whereas central venous thrombosis, precipitants in the catheter lumen, and mechanical obstruction do not respond (Box 87-1).

Precipitants within the catheter lumen most often result from flushing of the line with a heparin solution instead of saline after total parenteral nutrition. Heparin precipitates with total parenteral nutrition fluids. Clots within the catheter lumen usually result from failure to flush the line with a heparinized saline solution after blood aspiration.

A chest radiograph should be obtained in all patients with persistently occluded catheters to evaluate catheter position and integrity. The catheter tip should be positioned just above the right atrium. Persistent right atrial placement can cause perforation of this thin-walled heart chamber or result in a right atrial thrombosis. Comparison with previous radiographs may be necessary to ensure lack of movement or displacement. In patients with withdrawal occlusion but appropriate catheter position and without

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**Box 87-1**

**Differential Diagnosis of Occluded Chronic Indwelling Catheters**

<table>
<thead>
<tr>
<th>Complete Occlusion</th>
<th>Withdrawal Occlusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clot in catheter lumen*</td>
<td>Catheter against vessel wall</td>
</tr>
<tr>
<td>Precipitate in catheter lumen</td>
<td>Fibrin sheath*</td>
</tr>
<tr>
<td>Mechanical obstruction</td>
<td>Ball valve or mural thrombus*</td>
</tr>
<tr>
<td><strong>Intermittent Complete Occlusion</strong></td>
<td><strong>Subclavian vein thrombosis</strong></td>
</tr>
<tr>
<td>and Withdrawal Occlusion</td>
<td><strong>Pinch-off syndrome</strong></td>
</tr>
</tbody>
</table>

*Usually responds to low-dose intracatheter urokinase.*
In the pinch-off syndrome the catheter is intermittently obstructed during both administration and withdrawal of fluids, typically within 3 weeks after catheter placement. A chest radiograph demonstrates narrowing of the catheter lumen as it passes between the clavicle and the first rib. The catheter must be removed because of fragmentation or embolization if left in place.54

Because engorged collateral circulation or swelling in the affected extremity is not universally present with subclavian vein thrombosis, this diagnosis should be considered in all patients who are unresponsive to declotting attempts. Catheter removal with systemic heparinization or catheter maintenance with high-dose fibrinolytic therapy is a therapeutic option for subclavian vein thrombosis.52,55 Mechanical occlusion is rare and requires surgical catheter replacement. Because of variations in approach, early consultation is recommended in patients who have occluded central venous catheters (Fig. 87-6).

### Catheter Laceration

If an external catheter laceration or fracture occurs, the catheter should be clamped over tape distal to the laceration close to the chest wall. The catheter can be repaired as long as the damage is more than 4 cm from the chest wall. After clamping, as an interim measure, the next step is to insert a 14-gauge, 2-inch shielded intravenous catheter (Angiocath) into the catheter; remove the stylus; tape securely; and flush with heparin. The catheter can then be used while a repair kit is obtained.56

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**Figure 87-6.** Approach to occluded indwelling catheter.
Catheter-Related Infections

Catheter infections can be local or systemic. Local infections primarily involve the skin and subcutaneous tissues surrounding the exit site, with erythema, tenderness, and no clinical or laboratory evidence of sepsis. Skin organisms are primarily responsible for local infections, especially coagulase-negative staphylococci. Studies show that local infections do not require catheter removal and resolve with antimicrobial therapy alone.

The source of systemic infection in patients with Hickman-Broviac catheters may be difficult to localize, particularly in immunosuppressed patients. The most common sites of systemic infection in any patient with a central venous catheter, in decreasing order of frequency, are the urinary tract, the anorectal area, the upper respiratory tract, and the catheter. The most common organisms causing catheter infection are coagulase-negative staphylococci, S. aureus, and Candida albicans. In immunocompromised patients with Hickman-Broviac catheters, gram-positive organisms now are responsible for more cases of sepsis than gram-negative bacteria. Accordingly, initial empirical therapy should include an antistaphylococcal drug in addition to the usual gram-negative coverage. A good empirical regimen is vancomycin (1 g) and gentamicin (1 mg/kg intravenously [IV]). All patients who have a possible vascular access infection should have two blood culture samples drawn. Comparison of blood culture samples drawn simultaneously through the catheter and from a peripheral blood vessel may assist in determining whether the catheter is the source of infection. Infections that do not extend through the vessel wall (pericatheter infections) can be successfully treated without catheter removal. Catheter removal is mandatory in patients with continued positive blood culture results despite therapy and in those with vascular access infections caused by Candida species.

Catheter-related septic central venous thrombosis can progress through and around the vessel wall to cause a perivascular infection or abscess. This rare but devastating complication is associated with serious morbidity and a reported mortality rate as high as 83%. Because of the lack of specific clinical findings, the most prominent diagnostic feature is continued bacteremia after catheter removal. Diagnosis is confirmed by venography or CT.

Removal of the catheter, intravenous administration of antimicrobials, and anticoagulation constitute appropriate initial therapy. Surgical treatment with thrombectomy and possible abscess removal and resolve with antimicrobial therapy alone.

Figure 87-7. A, Groshong catheter with closed distal end and vocal cord-type valve. B, Hickman catheter with open distal end. (From Delmore JE, et al: Experience with Groshong long-term central venous catheter. Gynecol Oncol 34:216, 1989.)

Vascular Access for Hemodialysis

Quinton-Mahurkar Catheter

The Quinton-Mahurkar catheter provides immediate and short-term vascular access for hemodialysis. Its advantages include bedside placement and a functional life up to 18 months. This single, flexible, polyurethane cannula has two separate D-shaped channels, each connected by a molded Y piece to a color-coded external port (see Fig. 87-5B). To protect against a disconnected cap, each limb of the Y piece has an attached clamp. The Quinton-Mahurkar catheter is placed by the Seldinger technique, most often in the subclavian vein and less often in the femoral vein.

The complication rate from central venous catheter hemodialysis is near 30%, most commonly catheter-related infections and thrombosis.

A Quinton-Mahurkar catheter can be used to obtain blood samples. After blood withdrawal, the line should be flushed with more than 10 mL of normal saline solution, followed by 5000 units of heparin in 1 mL of saline to prevent intracatheter clot formation. The catheter can also be used for the administration of intravenous therapy. Routine care and use are otherwise similar to the care and use previously described for chronic indwelling central venous catheters.

Cimino-Brescia Fistula and Prosthetic Bridge Fistula

The subcutaneous Cimino-Brescia fistula is the preferred access for long-term hemodialysis. The fistula is created through a side-to-side and side-to-end anastomosis with use of the radial artery and the cephalic forearm vein. The high blood flow and pressure on the venous side of the fistula “arterialize” the veins in 3 to 5 weeks. The Cimino-Brescia fistula is well tolerated by patients, has a low infection rate, and has the longest functional
Thrombosis

Thrombosis is the most common complication of a subcutaneous arteriovenous fistula or prosthetic graft. Circumferential bandages, tourniquets, or blood pressure cuffs are avoided in the fistula-bearing arm because restricted venous outflow may predispose to thrombosis. The fistula can be used to acquire blood or vascular access (without use of a tourniquet). Normal graft flow is verified by feeling a thrill or hearing a bruit on auscultation. A strong palpable pulse with no matching thrill suggests venous outflow obstruction or early graft thrombosis. Thrombosis of arteriovenous fistulae necessitates temporary vascular access and usually the creation of a new fistula proximal to the thrombosed shunt.

Blood Withdrawal

Ideally, an alternative peripheral venipuncture site should be sought first before a Cimino-Brescia fistula is used for blood withdrawal. When an alternative site is unavailable, however, the arteriovenous fistula is a reasonable choice. An individual skilled in venipuncture techniques should maintain absolute sterility with antiseptic skin preparation, sterile gloves, and sterile gauze. Tourniquets are contraindicated and unnecessary. Venipuncture should be performed on the well-developed venous side of the fistula. After blood acquisition, gentle pressure should be maintained for 5 minutes, with care taken not to occlude the vessel lumen. The site should then be observed for several minutes to ensure that bleeding does not occur. A prosthetic arteriovenous bridge fistula can also be used to obtain blood samples with careful perforation of the superficial wall of the prosthetic graft; otherwise, the technique is identical.

Clinically differentiating a Cimino-Brescia fistula from a prosthetic arteriovenous bridge fistula may be difficult. The prosthetic portion of an arteriovenous bridge fistula connects the arterial to the venous vessels in an H shape and is tunneled for some distance beneath the skin, giving the appearance of a single large blood vessel. The prosthetic fistula has a thrill but is not as pulsatile as a Cimino-Brescia fistula when gently palpated. If asked, most patients are knowledgeable about their fistula.

Peripheral intravenous access is best established at an alternative site. When an alternative site is unavailable and the patient requires timely intravenous access, the Cimino-Brescia fistula or bridge fistula can be used, following the guidelines for venipuncture. Careful attention to sterile technique, operator skill, and avoidance of tourniquets can provide timely venipuncture or intravenous therapy while preventing infectious or thrombotic complications. If an intravenous line is used in a fistula, it should be replaced by alternative intravenous access as soon as possible.

Infection

Infections of an arteriovenous fistula or graft are potentially life-threatening and manifest with signs of septicemia and local inflammation. Once the diagnosis of infected fistula is considered, blood cultures should be obtained and intravenous antibiotics for gram-positive skin organisms administered. Prosthetic graft infection cannot be eradicated with intravenous antibiotics alone and requires prosthetic graft removal. Infections are the second leading cause of death of patients undergoing long-term dialysis.

Steal Phenomenon

Vascular steal from the ulnar artery via the palmar arch occasionally occurs in patients with atherosclerotic disease distal to the shunt, particularly in diabetic patients. Symptoms of fingertip ischemia occur during periods of increased shunting (hemodialysis or increased activity). The steal phenomenon usually requires graft ligation with construction of a new fistula in the opposite extremity.

Venous Hypertension

Acute venous hypertension is a true surgical emergency that occurs in the first few weeks after fistula construction. The early rise in venous pressure produces marked swelling of the extremity, severe venous stasis and characteristic skin pigmentation, edema, and occasionally venous ulceration. Management of venous hypertension requires hospitalization and urgent ligation of the vein immediately distal to the fistula before a potentially exsanguinating vessel rupture.

Bleeding

Patients also are seen in the emergency department with bleeding from their fistula after dialysis. Persistent, gentle pressure, with care taken not to occlude blood flow, usually resolves this problem.

The references for this chapter can be found online by accessing the accompanying Expert Consult website.
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