Basic Vascular Pathophysiology of the Hand, Wrist, and Forearm

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Vascular disease of the upper extremity consists of a diverse group of conditions of different etiologies and presentations. These conditions can be broken down into the following groups: (a) vaso-occlusive disease, which consists of embolism, thrombosis, aneurysms, atherosclerosis, and vasculitis; (b) vasospastic conditions, including Raynaud’s disease, secondary Raynaud’s phenomenon, acrocyanosis, erythromelalgia, and livedo reticularis; (c) compressive vascular disease; (d) congenital vascular disease; (e) congenital vascular malformations, which include hemangioma and vascular malformation; (f) vascular tumors; and (g) venous occlusive diseases.

VASO-OCCLUSIVE DISEASE

Embolism

Embolism is a common vascular disease of the upper extremity. Sudden onset of pain, paresthesia, paralysis, and loss of distal pulses herald an onset of embolus to the upper extremity vessels. Of all arterial embolizations, 10% to 20% involve the upper extremity. Two-thirds of those that involve the upper extremity originate from a diseased heart, usually from fibrillation. A small percentage follow acute myocardial infarction, apparent or even silent. Other sources of arterial emboli include (a) a thoracic outlet with poststenotic dilatation, (b) aortic and ventricular aneurysms, (c) atheromatous plaques, (d) sequelae of cardiac valve replacement, and (e) foreign bodies. Compared to arterial thrombi, those emboli that originate in the heart lodge in more proximal vessels—for example, bifurcation of the brachial artery.

The most frequent signs of an embolus are (a) altered limb color, (b) decreased temperature, (c) absent peripheral pulses, (d) sensory changes, (e) pain, and (f) loss of some or all motor function. The point of occlusion can be determined by palpating along the course of the larger vessel to the point at which pulsations cease. A Doppler flowmeter can usually demonstrate the level of occlusion in smaller vessels. Significant sensory changes or peripheral paralysis may be signs of advanced ischemia and warn of impending gangrene.

Because the surgical procedure to be performed is dictated by the type of occlusion, differentiation between embolism and thrombosis is important. Collateral circulation does not develop distal to emboli, as it so often does in chronic arterial sclerotic occlusion; hence, the distal ischemia is more severe, as evidenced by pallor, pain, coldness, and complete absence of peripheral pulses. When in doubt, arteriography or magnetic resonance angiography is indicated and enables the physician to (a) establish the diagnosis, (b) establish the level and completeness of the main channel occlusion, (c) determine the type of lesion (e.g., underlying arterial venous fistula or aneurysm), and (d) evaluate collateral circulation and, in patients with preexisting arterial sclerotic disease, the presence of distal runoff (Fig. 1).

Surgical embolectomy is carried out via an arteriotomy at the point of cessation of arterial pulsations. After removal of the embolus, proximal and distal blood flow should be normal. If not, Fogarty catheters are inserted to make sure that all clots have been removed. If there is any doubt about adequate distal perfusion of the extremity, an operative arteriogram can be done or distal incisions can be made, and the vessel in question can be explored with small-caliber Fogarty catheters. Postoperative anticoagulation is indicated. Oral anticoagulation with warfarin sodium (Coumadin) is continued for 3 months postoperatively.

With embolisms that are distal to the wrist, initial efforts are directed at trying to reverse the vasospastic component of ischemia with stellate ganglion or axillary anesthesia. Heparin is also started. In the absence of response to these measures, thrombolysis with intraarterial or intravenous urokinase is tried. If the arterial tree reclogs, one must be suspicious that some of the clot remained or that underlying venous occlusion or disease exists. If significant soft tissue swelling occurs from prolonged muscle ischemia, venous drainage may be impeded. If permitted to go unchecked, this would finally occlude not only venous, but also arterial, channels. When such occlusion is suspected or anticipated, fasciotomy is indicated.
Arterial thrombosis may be acute, as is seen in trauma, or chronic, from underlying vascular disease. Arteriosclerotic thrombosis is much less common in the upper extremity than in the lower. It often occurs at or near the bifurcation of a large arterial branch. In contrast to embolism, however, any arterial segment may be involved.

Careful history reveals symptoms of chronic ischemia before the final closure of the narrow lumen by the thrombus, which results in acute symptoms. Proximal to the thrombus, the pulses may be weak, and there may be a bruit. Doppler examination confirms the point of occlusion. Noninvasive evaluation determines the overall status of the digital blood flow and the adequacy of flow in the ischemic portion of the hand. A brachial pressure difference of more than 20 mm Hg suggests proximal disease. McNamara et al. (1) noted proximal disease in 22 patients who had significant differences in proximal and distal brachial pressures. When proximal disease is suspected, arteriography is helpful in establishing the exact nature and location of the disease. In patients with more distal disease, such as ulnar artery occlusion, assessment with noninvasive techniques such as pulse-volume recording (PVR) and Doppler examination is often adequate. If there is clinical evidence of proximal disease, however, invasive techniques may be required.

In our experience, patients with ulnar artery occlusion who have excellent flow, as demonstrated by noninvasive studies and normal digital pressures in all digits, usually do not require reconstruction of the thrombosed vessel at the wrist level. Zimmerman et al. (2) confirm our concept. An exception is the individual with emboli that are secondary to thrombotic occlusion at the wrist level.

Nonoperative treatment may be used when patients have less marginal arterial flow or are poor risks for surgical intervention. These techniques include (a) horizontal positioning of the extremity at room temperature; (b) anticoagulant therapy, such as aspirin, low-molecular-weight dextran, heparin, or warfarin sodium; (c) adequate sedation and analgesia; (d) sympathetic block; (e) vasodilating drugs (e.g., calcium channel blockers, such as nifedipine); (f) medication that affects red-cell flow characteristics, such as pentoxifylline (Trental); and (g) intraarterial fibrinolytic therapy. Nifedipine, in our experience, is more effective in vasospastic conditions than in vaso-occlusive disorders.

Surgical correction of arteriosclerotic occlusion or thrombosis is indicated when there is significant diminution in flow or when viability of the extremity is in question (Figs. 7, 3, and 4). Excision of the thrombosed segment and reconstitution with a reversed vein graft using microsurgical technique is indicated. Arterial grafts also give good results; however, donor sites are less readily available.

McGregor et al. (3) have shown that blood flow and skin temperature of the involved upper extremity increase in patients under brachial plexus anesthesia. These authors suggest that such anesthesia therefore may be the procedure of choice in arterial reconstruction.

Intraarterial fibrinolytic therapy has been used to reestablish blood flow in the presence of occlusion that is secondary to thrombosis or embolism. Streptokinase and urokinase have been compared in this regard by Belkin et al. (4). Urokinase had a significantly higher success rate and fewer complications than streptokinase, whereas the primary advantage of streptokinase was its lower cost. Belkin et al. (4) concluded that the increased efficacy and safety of urokinase made it the agent of choice for intraarterial fibrinolytic therapy. Thrombolytic therapy as such is generally used before but not after surgical intervention due to the increased bleeding risk once surgery has been performed.
Ulnar Artery Thrombosis

Thrombosis of the ulnar artery was first described by von Rosen in 1934 (5). The patient was a 23-year-old factory worker, who struck the edge of a metal bar with the palm of his hand and experienced acute pain and swelling. After 2 weeks, he also had signs and symptoms of hand ischemia. The distal ulnar artery was explored, and the thrombosed part was excised, resulting in relief.

Various names have been given to this condition, such as hypothenar hammer syndrome (6), posttraumatic digital ischemia, and pneumatic tool disease. It occurs in people who use the palm of their hands during daily activities, such as pushing, hammering, pounding, or twisting.

According to Koman and Urbaniak (7), the ulnar artery is most vulnerable in the distal part of the Guyon’s canal, where the protective effects of the palmaris brevis is lost. Repeated trauma to the hypothenar area causes periadventitial scarring, which results in damage to the media, disruption of the internal elastic lamina, intimal damage, and subintimal hematomas (Fig. 5) (8). The thrombosis may extend subsequently to the common digital arteries and even to the digital arteries (Fig. 6). Embolization can also take place, and showers of emboli can occlude flow downstream in the digital vessels. Anatomic peculiarities of the blood vessels of the hand could also explain the genesis of symptoms in this condition. In 21.5% of hands, the superficial palmar arch is incomplete. These patients are more susceptible and exhibit symptoms of ulnar artery thrombosis. Structural change in the artery can give rise to vasospasm and further reduction in the local blood flow. The vasospasm may extend well beyond the confines of the thrombosed segment. The excision of this segment tends to result in cessation of the vasospastic effect (Fig. 7).
FIGURE 4. Long-segment thrombosis of the ulnar artery (close-up).

FIGURE 5. Angiogram that demonstrates blockage of the ulnar artery.

FIGURE 6. Angiogram that shows decreased circulation in the ulnar digits of the hand.
Patients are typically males who are in their fourth decade of life, with 60% of the patients presenting with a history of repeated trauma. Unilateral involvement is common. Pain is usually the universal complaint. The pain is usually localized in the ulnar digits. In the presence of significant arterial obstruction, night pain or even rest pain may be a troublesome symptom.

Cold intolerance presents in 80% of the population and is often a disabling symptom. In Koman and Urbaniak's series (7), moderate or severe symptoms of cold intolerance correlated well with incomplete circulation and had abnormal temperature responses, whereas mild symptoms had normal cold stress testing.

Numbness in the sensory distribution of the ulnar nerve occurs in approximately three-quarters of this patient population, with more than one-half demonstrating a positive Tinel's sign at the Guyon's canal level. Signs of peripheral nerve irritation are present in the ulnar nerve distribution in the form of increased static and moving two-point discrimination and motor weakness of intrinsics.

In a series by Conn et al. (6), of the 13 hands that presented signs of ischemia, the ring finger was involved in six instances, the index finger in four, the small finger in three, and the long finger in two.

Investigation of cases of suspected ulnar artery thrombosis include thermography and temperature probes, Doppler mapping, real-time ultrasonography, radionuclide scanning, color flow Doppler imaging, magnetic resonance angiography, and traditional arteriography.

The goal of treatment is to increase arterial inflow to the digits. Arterial resection removes the stimulus for sympathetically driven, increased vascular tone, whereas arterial reconstruction adds more inflow to the hand. Mehlhoff and Wood (9) found that 88% of their vein grafts were patent at 1 year, as demonstrated by Doppler flow and Allen's tests. They found that functional results improved in 57% only if the grafts remained patent. In their series, the favorable outcomes were in (a) nonsmokers, (b) patients with a single distinct history of trauma to the wrist, and (c) patients with less than 5 months of history of symptoms. Zimmerman et al. (2) have shown that intraoperative vascular monitoring is used to guide the decision between reconstruction and resection. After arterial resection, intraoperative digital pressures are obtained. If the digital brachial index (the finger pressure divided by the brachial artery pressure) is less than or equal to 0.7, flow to the digit is inadequate, and reconstruction is required. If the digital brachial index is greater than 0.7, flow to the digits is adequate, and reconstruction is superfluous. In the event that intraoperative pressure monitoring is unavailable, rapidity of refill time after tourniquet deflation is used as a guide for reconstruction. If the refill is rapid (less than 6 seconds), reconstruction is deemed unnecessary (2).

Radial Artery Thrombosis
Most cases of radial artery thrombosis are due to trauma or occur as a result of the use of percutaneous indwelling radial artery catheters (10, 11). In this area, the artery is subjected to repetitive injury, as well as damage, by the tendons of the first dorsal compartment. Ruch et al. (12) described 13 cases of radial artery reconstruction through the use of a reversed interpositional vein graft. The authors point out that all vessels were thrombosed as they emerged from the protection of the brachioradialis and passed under the first dorsal compartment.

Digital Artery Thrombosis
Lowrey et al. (13) studied digital vessel flow in professional baseball catchers by using modified Allen's test and the Doppler flow test. They also studied perfusion of the hand by using radioactive technetium. Thirteen out of 22 baseball catchers had evidence of decreased circulation in the left index finger. They suggested the addition of padding to the glove.

Medial Artery Thrombosis
Median artery thrombosis is rare but can present as carpal tunnel syndrome.

Thrombosis in Immune Disorders
Certain autoimmune diseases, particularly systemic lupus erythematosus (SLE), can present with extremity thrombosis.
These effects are due to the following mechanisms:

Abnormalities of the complement system. This occurs in acquired angioneurotic edema in which there is a protein C1 deficiency. Normally, protein C1 prevents platelet binding to collagen. In a protein C1 deficiency, platelet aggregation is induced by collagen, thus causing clotting. It also occurs in paroxysmal nocturnal hemoglobinuria. Platelets in this condition can fix larger amounts of protein C3 than normal platelets. Platelet aggregation is triggered by activation of alternative complement pathways.

Cell-mediated attack on the vessel wall. Examples are (a) Behçet’s syndrome and (b) thromboangiitis obliterans.

Specific humoral antibodies. Lupus anticoagulant, an antibody (immunoglobulin G or M, or both), is sometimes found in patients with SLE. Thirty percent to 50% of SLE patients with the lupus anticoagulant exhibit one or several episodes of thrombosis, whereas thrombosis is rare in the SLE patient without the lupus anticoagulant.

One of the following mechanisms is thought to play a role in the action of the lupus anticoagulant in the production of thrombosis:

Affinity of the lupus anticoagulant (which is negatively charged) for phospholipids interferes with the availability of arachidonic acid and the release of prostacyclin (which is normally stored within phospholipids).

Interference with antithrombin III activity.

Inhibition of thrombomodulin. Thrombomodulin is an endothelial cofactor, which helps in the conversion of thrombin to protein C. Thus, feedback inhibition of coagulation by activated protein C is impeded.

Defective intravascular fibrin removal.

Aneurysms

In a large series during the Korean War, although more than 45% of aneurysms involved the lower extremity, only 3.8% were in the upper extremity (14). False aneurysms of the upper extremity accounted for almost one-third of all false aneurysms in the Vietnam Vascular Registry (15).

FIGURE 8. Exploration of first web swelling reveals a radial artery mass.
True and false aneurysms occur in the upper extremity. A false aneurysm generally arises after trauma, when the arterial wall is disrupted, and blood flows into the surrounding tissue (Figs. 8, 9, 10). The resulting acute, pulsatile hematoma is contained by the surrounding tissue. Fibroblasts then proliferate and form a thick false capsule (16). Characteristically, the false aneurysm has no elastic or muscle fibers in its wall. The traumatic false aneurysm is the most common aneurysm of the hand.

A true aneurysm is a space or sack that is formed by the widening of the lumen of an artery (Fig. 11). Histologic findings of arterial wall elements (muscle and elastic fibers) in the aneurysm wall confirm the diagnosis of a true aneurysm (17) (Fig. 12).

True aneurysms may be subdivided into idiopathic, arteriosclerotic, traumatic, and mycotic types. Traumatic aneurysm is the most frequent type of true aneurysm found in the hand and can be subclassified into acute traumatic and chronic occupational palmar aneurysms (18).
Aneurysms present as gradually expanding masses that are usually painless, unless they are associated with vascular insufficiency or pressure against an adjacent nerve. Pressure on an adjacent nerve can result in paresthesias or hyperesthesia. Distal thrombosis is associated with cyanosis, coolness of the part, and a prominent pulsation. Raynaud's phenomenon, trophic changes, and distal necrosis may follow decreased flow or embolization. Larger, more proximal aneurysms have an associated thrill or bruit. An aneurysm rarely presents with an acute rupture.

Kleinert et al. (19) found that most aneurysms in the hand occur after trauma to the radial or ulnar artery, where they course unprotected by fascia across the unyielding hook of hamate and the prominence of the trapezium. These authors were the first to report vascular reconstruction for aneurysms of the hand. In that paper, they report on 11 aneurysms: five ulnar, four radial, one posterior interosseous, and one perineural artery.

Ulnar artery aneurysms are more common than other aneurysms in the hand (Figs. 13, 14, 15 and 16). These aneurysms occur in the unprotected part of the ulnar artery distal to the Guyon's canal to the point where it is covered by the palmar fascia. In this short vulnerable segment, the ulnar artery is covered by skin, subcutaneous tissue, and the few fibers of the palmaris brevis muscle and rests on the hamate. Repeated blunt trauma to the hypothenar eminence by using this part of the hand as a hammer has resulted in the term hypothenar hammer syndrome (6).
Rothkopf et al. (20) reported on ten ulnar artery aneurysms. Blunt trauma was responsible for 70% of their cases, whereas penetrating trauma resulted in aneurysm formation in 20%. There was no history of trauma in the remaining 10% of cases. Three of their ten cases that presented with asymptomatic masses of a duration of 4 weeks or shorter had no evidence of emboli. However, seven cases that presented with vascular symptoms of a duration of 6 weeks or longer had definite signs of digital embolization of which ring and long fingers were the most common sites.
Ulnar artery aneurysm may present as a pulsatile mass, along with ischemic changes in the form of cold intolerance, pain, and mottling (Figs. 17 and 18). If digital embolization occurs, subungal hemorrhage, ulceration, or gangrene can occur. Compression of the digital nerve produces signs of numbness, paresthesia, and pain, with a positive Tinel's sign and decreased two-point discrimination. Bone erosion may be present in rare circumstances.

Radial artery aneurysms occur on the distal forearm along the dorsal route of the radial artery over the scaphoid edge of the trapezium and first metacarpal, where the artery rapidly dips between the heads of the first dorsal interosseous muscle.

Digital artery aneurysms are relatively rare. Few cases of false aneurysms have been reported. Layman et al. (21) reported his first case of true aneurysm of a digital artery. Subsequently, Ho et al. (22) added four more cases. Both of these authors and others have speculated on the rarity of digital aneurysms, given the predilection of the digits to injury. The small caliber of the digital artery would not favor a partial arterial wall injury, which is a prerequisite for false aneurysms. Similarly, blunt trauma is more likely to damage the intima, resulting in thrombosis, rather than the media, which could give rise to a true aneurysm.
FIGURE 17. Exploration of the ulnar artery shows the aneurysm.

Noninvasive techniques allow assessment of flow that is distal to the aneurysm to determine if blood supply is adequate or diminished. Because aneurysms at, and distal to, the wrist level are often easy to examine, arteriography is usually not necessary. Aneurysms that are proximal to the wrist are more difficult to delineate clinically because of the surrounding muscle mass; under these circumstances, the physician may wish to use arteriography and ultrasound as primary diagnostic tools. Proximal embolic sources must be ruled out with arteriography from the aortic arch to the digital tips (23). PVR should be correlated with the arteriogram, especially in the peripheral embolism.

The preferred treatment for nonmycotic aneurysms is resection followed by direct anastomosis or vein graft.

FIGURE 18. Clinical photograph that shows ischemia changes of ring and small fingers.

Mycotic aneurysms may be divided into primary and secondary types. Primary mycotic aneurysms occur in the absence of overt sepsis, whereas secondary mycotic aneurysms result from intravascular infection and direct extension of a septic focus into an adjacent blood vessel. Drug addiction has increased the number of mycotic aneurysms in the brachial artery. Other etiologies include vascular reconstruction procedures, hemodialysis shunts, urinary tract infections, gonorrhea, osteomyelitis, and penetrating trauma. The most common offending associated organisms are *Staphylococcus aureus* and *Streptococcus*. Others include enterococcus, salmonella, gonococcus, and clostridia.

The classic sign of a mycotic aneurysm is a fever that is associated with a painful, tender, pulsatile mass that is adjacent to a blood vessel. Other signs of true aneurysm may also be present.

The treatment of choice for mycotic aneurysms is excision and ligation followed by at least 6 weeks of antibiotics. If the extremity appears ischemic after excision and ligation, an extraanatomic bypass with an autogenous vein graft should be performed away from the site of excision.

Atherosclerosis

Atherosclerotic plaques that cause upper extremity problems are most commonly located in the subclavian and axillary arteries. Patients with occlusion of the subclavian artery that is proximal to the origin of the vertebral artery may present with a *subclavian steal syndrome*. Reduction in blood pressure distal to the occlusion causes blood to flow...
retrograde through the vertebral artery. This was first described by Harrison in 1829 (24) but was expanded on by Fisher (25).

Blood now flows from the cerebral circulation through the vertebral system to augment the supply of the ischemic arm (26). In this patient population, general atheromatous lesions are common, especially those that involve the cerebral circulation that manifest in the classic symptoms of dizziness or vertigo or repetitive use of the arm. In one series, 88% of symptomatic patients had central nervous system symptoms, and 40% had intermittent claudication in the upper extremity. There is a marked predilection for the left side to be involved (with a ratio of 3:1) (27).

Occlusion of the subclavian artery distal to the axillary artery may be asymptomatic owing to the rich collateral circulation. The distribution is equal on both sides. In symptomatic patients, intermittent claudication is most frequent.

Brachial artery occlusion is usually due to trauma or cardiac catheterization. The general rate of surgical intervention after cardiac catheterization is approximately 1%. Ischemic symptoms usually become evident when patients resume normal use of their hands.

Obliterative disease of the arteries of the forearm, hand, and fingers is usually caused by distal embolism from proximal atheromatous plaques.

Proximal occlusion can be detected by noninvasive vascular studies that include bilateral brachial pressures. Pressure differences of 40 to 50 mm Hg occur in patients with symptoms of intermittent claudication and ischias. On the other hand, patients with subclavian steal syndrome who exhibit central nervous system symptoms have less of a pressure differential (20 to 40 mm Hg) due to shunting of blood through the vertebral artery. One of the classic signs of proximal compression is a bruit over the subclavian artery.

A definite diagnosis, along with demonstration of the lesion, is done by angiography. Percutaneous transluminal angioplasty should be attempted with anticoagulant cover. If this is successful, anticoagulation is continued for a period of 3 months.

Surgical options currently include extraanatomic bypass grafts with vein or prosthetic material. These include subclavian to carotid, subclavian to subclavian, or axillary to axillary bypass grafts.

**Vasculitis**

Vasculitis of varying etiologies can be an important cause of occlusive disorders of the upper extremity. The presentation of digital ischemia should initiate a search for an underlying pathologic condition that precipitated the process of vasculitis (Table 1).

**Causes of Vasculitis**

*Thromboangiitis obliterans*, also known as *Buerger’s disease*, is a vasculitis involving medium-sized and small arteries and veins that begins in distal vessels in the extremities and then progresses proximally in a skip-zone manner (28). Buerger’s disease should be suspected in patients of any nationality who are between the ages of 20 and 50 years who present with signs and symptoms of intermittent claudication, chronic recurrent sepsis, rest pain, or migrating phlebitis. There is a high degree of association with tobacco users. Tobacco use is a trigger for an autoimmune mechanism in a group of patients. Withdrawal of tobacco prevents tissue damage. Upper extremity involvement occurs in 15% to 20% of patients. Migratory thrombophlebitis occurs not infrequently as a precursor, followed by aching or burning pain, which is most intense in muscle compartments. The pain is disproportionately severe to the objective evidence of ischemia—a slight bluish discoloration and some surface coolness. Common signs are muscle atrophy, skin and digital pulp atrophic changes, ulceration, edema, and absent peripheral pulses (Figs. 19 and 20).

Conservative management has a definite place in the overall treatment. Of greatest importance is the discontinuation of smoking. Other advocated measures include the administration of corticosteroids during episodes of acute exacerbation, the administration of anticoagulants and antibiotics, the practice of Buerger’s exercises, the practice of local hygiene, and the use of protective dressings.

### TABLE 1. CHARACTERISTICS OF VASCULAR BIRTHMARKS

<table>
<thead>
<tr>
<th>Hemangioma</th>
<th>Malformation</th>
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<tbody>
<tr>
<td><strong>Clinical</strong></td>
<td></td>
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<tr>
<td>Usually nothing seen at birth; 30% of cases present as red macule</td>
<td>All present at birth; may not be evident</td>
</tr>
<tr>
<td>Rapid postnatal proliferation and slow involution</td>
<td>Commensurate growth; may expand as a result of trauma, sepsis, and hormonal modulation</td>
</tr>
<tr>
<td>Female to male ratio is 3:1</td>
<td>Female to male ratio is 1:1</td>
</tr>
<tr>
<td><strong>Cellular</strong></td>
<td></td>
</tr>
<tr>
<td>Plump endothelium, increased turnover</td>
<td>Flat endothelium, slow turnover</td>
</tr>
<tr>
<td>Hematologic</td>
<td>Radiologic</td>
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<tr>
<td>-------------</td>
<td>------------</td>
</tr>
<tr>
<td>Increased mast cells</td>
<td>Multilaminated basement membrane</td>
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<tr>
<td>Normal mast cell count</td>
<td>Normal thin basement membrane</td>
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**Hematologic**

- **Primary platelet trapping; thrombocytopenia (Kasabach-Merritt syndrome)**
- **Primary stasis (venous); localized consumptive coagulopathy**

**Radiologic**

- **Angiographic findings: well-circumscribed, intense lobular-parenchymal staining with equatorial vessels**
  - Angiographic findings: diffuse, no parenchyma
  - Low flow: phleboliths, ectatic channels
  - High flow: enlarged, tortuous arteries with arteriovenous shunting

- **Infrequent mass effect on adjacent bone; hypertrophy rare**
- **Low flow: distortion, hypertrophy, or hypoplasia**
- **High flow: destruction, distortion, or hypertrophy**


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**FIGURE 19.** Clinical photograph of a patient with Buerger’s disease that shows ulceration of index and thumb.

Surgical treatment includes sympathectomy; amputation, as indicated; and, occasionally, vascular reconstruction. The patient with nearly normal-appearing skin that is cool, moist, and, perhaps, a bit cyanotic is expected to have improvement from sympathetic denervation, whereas the patient with dry, atrophic skin that has loss of substance is probably not expected to have improvement. One preoperative test to determine the value of sympathectomy is the stellate ganglion block followed by observation of the patient’s objective and subjective response. Cold stress testing before and after blockade can be performed to determine the change in blood flow.

Sympathectomy in Buerger’s disease has the potential benefits of (a) subjective improvement, (b) decrease in severity of vasospastic episodes, (c) possible improvement in collateral circulation, and (d) help in preventing amputation. In the past, the transaxillary approach for sympa-thectomy was preferred. Digital sympathectomy has been clinically effective after unsuccessful transaxillary sympathectomy in patients with Buerger’s disease.
**Hypersensitivity Vasculitis**

Antigens that cause hypersensitivity vasculitis can be a drug (sulfa drugs), a bacteria (β-hemolytic streptococcus), a tumor antigen, or a serum protein. Allergic or hypersensitivity vasculitis affects small vessels, especially postcapillary venules, and has a good prognosis (29). The first step in management is the withdrawal of the antigen and the administration of an antiinflammatory medication, such as prednisone. Causes of hypersensitivity vasculitis include serum sickness, Schönlein-Henoch purpura, and malignancies.

**FIGURE 20.** Angiogram of a patient with Buerger’s disease (the same patient who is shown in Fig. 19) that shows decreased circulation of index and thumb.

**Collagen Diseases**

Antigen antibody complex deposition in digital vessels is the common factor in this group of heterogeneous conditions. Following these changes, fibrinoid degeneration, intimal thickening, and superimposed vasospasm produce the clinical features of digital ischemia.

**Rheumatoid Arthritis**

Seropositive rheumatoid arthritis gives rise to obliterative endarteritis in the digital arteries. Postcapillary venules in the skin are also affected. Generally, severely affected hands and digits are subject to vasculitis. A careful search for vasculitis must be made before surgical reconstruction of these deformed hands.

**Systemic Lupus Erythematosus**

Vasculitis accounts for the Raynaud’s phenomenon of SLE, which occurs in one-fifth of SLE patients. Digital ischemia may indeed be the presenting symptom of SLE and predates arthritic symptoms. Arteriolar endothelial destruction with eosinophilic infiltration of the intima of the common digital arteries narrows the lumen.

**Scleroderma**

Patients with CREST syndrome (calcinosis cutis, Raynaud’s phenomenon, esophageal motility disorder, sclerodactyly, and telangiectasia) develop vasculitis and digital ischemia quite frequently after predating systemic disease by many years. The arterial involvement occurs in digital arteries, especially those of the long and ring fingers. The process also involves the ulnar artery in 50% of cases. Pathologically, there is intimal thickening secondary to fibrin deposition in the vessel wall, which results in narrowed arterial lumen and digital ischemia.

**Polyarteritis Nodosa**

Polyarteritis nodosa involves small and medium-sized vessels with segmental involvement.

**Blood Dyscrasias**

Cold agglutination, cryoglobulinemia, and polycythemia vera can present with vasculitis.

**Drug-Induced Vasculitis**

Ergotamine that is used for the treatment of migraine can result in vasoconstriction due to its direct effect on the beta receptor, which results in long segments of smooth muscle spasms.

**VASOSPASTIC DISORDERS**

Raynaud’s disease or primary Raynaud’s phenomenon (Fig. 21) occurs in the absence of underlying predisposing
systemic abnormalities. It is therefore a disease based on the exclusion of other etiologies. Certain features are helpful in arriving at the diagnosis: (a) 80% of patients are women; (b) the disease is present in other family members; (c) the age of onset is between 20 and 30 years of age; (d) the fingers are involved first, toes later; (e) minor ulcerations, chronic paronychias, and small areas of necrosis, which rarely lead to major amputations, are present; (f) numbness and color changes are precipitated by cold or emotional stress; (g) involvement of all fingers, exclusive of the thumb, is bilateral and symmetric; (h) organic arterial disease is absent (it is particularly important to consider Buerger’s disease in men); (i) other conditions that might cause Raynaud’s phenomenon are absent; and (j) the disease must have been present for 2 years or longer. Allen and Brown (32) established these features as necessary for the diagnosis of Raynaud’s disease, because many underlying occult collagen diseases take time to become manifest.

**Secondary Raynaud’s phenomenon or Raynaud’s syndrome** is a vasospastic disorder that is characterized by transient, intermittent attacks of asymmetrically located vasospasm that involves one or several digits and rarely lasts longer than a few minutes. The attacks are precipitated by cold or emotional stress. Usually, there is a sequence of color changes, that is, initial pallor followed by cyanosis or ruber, which is associated with burning pain or paresthesia, before returning to normal skin color. The subsequent signs and symptoms depend on whether the vasospasm of the arteries and veins, which produces the initial pallor, is relieved simultaneously or if one is relieved before the other.

Secondary Raynaud’s phenomenon, as its name implies, is secondary to some underlying pathology. It behooves the physician to determine the more obscure problem (30), which may be (a) occlusive arterial disease (arteriosclerosis obliterans, thromboangiitis obliterans, or arterial occlusion due to thrombosis or embolism); (b) a collagen disease, including rheumatoid arthritis, SLE, polyarteritis nodosa (31,32), scleroderma, dermatomyositis, or mixed connective tissue disease; (c) recurrent trauma to the hand followed by thrombosis or aneurysm, which occurs in occupations, such as stonecutter, pneumatic tool operator, mechanic, farmer, and butcher; (d) various neurologic abnormalities, such as nerve compression, peripheral neuritis, multiple sclerosis, progressive muscular dystrophy, causalgia, and glomus tumors; (e) thoracic outlet syndrome (TOS); (f) heavy metal intoxication; (g) a drug-induced condition (31), as through the use of ergot, beta-blockers, cytotoxic drugs, and oral contraceptives; (h) previous cold injury (frostbite); (i) cryoglobulinemia, which is suspect when vasomotor phenomena are severe enough to affect exposed areas, such as the nose, ears, and lips; and (j) a metastatic or multiple myeloma, which can produce cryoglobulins.

Rivers and Porter (31) noted that by far the most common connective tissue disease in patients with secondary Raynaud’s phenomenon is scleroderma. One-third to one-half of patients with Raynaud’s syndrome in several reviews have scleroderma. Likewise, Raynaud’s syndrome was found in 70% to 90% of scleroderma patients. These authors also reported that 85% of patients with mixed connective tissue disease, 25% of patients with rheumatoid arthritis, and 20% of patients with SLE have Raynaud’s syndrome.

The requirement for a diagnosis of Raynaud’s syndrome is episodic color changes in response to cold or emotional stimuli. Further diagnostic testing may be done. The authors measure temperature of the hand before and after its immersion for 3 minutes in water at 15°C. Most patients with Raynaud’s syndrome take longer than 20 minutes for hand temperatures to return to normal.

These patients with abnormal cold recovery undergo laboratory studies including complete blood cell count, erythrocyte sedimentation rate, and detection of rheumatoid factor and antinuclear antigens. Other tests that may be of value are urinalysis serum protein electrophoresis, antibodies to nuclear antigens, antinuclear DNA and extractable nuclear antigen, determination of C3 complement level, and immunoglobulin electrophoresis (31). Radiographic evaluation of the hands is also obtained. The finding of calcinosis in the distal phalanges, along with history of cold intolerance, is usually consistent with the presence of scleroderma.

Patients with cold intolerance may have underlying vaso-occlusive disease as a primary problem instead of vasospastic disease. Doppler examination and PVR aid in detection of underlying vaso-occlusive disease. Sumner and Strandness (33) identified a peaked waveform and chronic notch in the photoplethysmography tracings of patients with Raynaud’s syndrome. Rivers and Porter (31), however, did not consistently record that finding in their patients.

Holmgren et al. (34) reviewed over 300 patients with Raynaud’s phenomenon; 70% had a significant associated disease process, whereas 30% did not. These authors essentially abandoned hand arteriography because of the reliability of digital plethysmography in differentiating spastic from obstructive disease processes. They also cautioned that a normal plethysmographic tracing could be present in a documented systemic disorder, such as Raynaud’s phenomenon.

Furthermore, these authors noted that Raynaud’s phenomenon existed in two distinct forms. The first form was a
spastic type that produced a distinctly abnormal digital artery constrictive response with cold exposure and was usually present in all fingers equally. This first form was seen in patients with primary or secondary Raynaud’s phenomenon. The second form was obstructive and appeared to represent a normal vasoconstrictive response to cold exposure. This form, which was found only in patients with Raynaud’s syndrome, often spared certain fingers (Fig. 22). It appeared secondary to arteriosclerosis, trauma, and end-stage autoimmune-type diseases in which severe vasospasm eventually led to actual digital vessel obstruction.

Treatment for primary and secondary Raynaud’s phenomenon consists first of minimizing cold exposure, wearing warm gloves, and avoiding tobacco. Some medications may aggravate cold intolerance.

Nifedipine has achieved recent popularity in treatment of Raynaud’s phenomenon. Our experience in a large number of patients is that 10 mg of nifedipine, three times a day, may be helpful. Other authors have reported similar success with nifedipine treatment (31, 35, 36, and 37). Hurst et al. (38) recommended a single intraarterial injection of 1.25 mg of reserpine into the brachial artery for a prolonged sympathectomy in treatment of reversible vascular spasm. This produced a chemical sympathectomy with prolonged vasodilatation that often lasted as long as 4 days. Finally, biofeedback has been used successfully in the treatment of some vasospastic conditions, including those that cause Raynaud’s syndrome.

**FIGURE 22.** Angiogram of end-stage Raynaud’s disease with extremely poor peripheral circulation.
Treatment of persistent digital vasospasm by digital artery sympathectomy was described initially by Flatt (39). A periarterial sympathectomy was performed at the origin of the proper digital arteries through stripping of the adventitia (Fig. 23). Wilgis (40,41) also described digital artery sympathectomy and recommended extending it to the proximal phalanx. Preoperative evaluation of patients who could benefit from sympathectomy can be determined in part by cold exposure testing followed by repeat testing under metacarpal block anesthesia. The return to normal recovery time with cold stress after metacarpal block suggested the patients were surgical candidates for digital sympathectomy. Some patients did have a beneficial response to digital sympathectomy after a failed transaxillary sympathectomy. Digital sympathectomy is an exacting procedure and must be performed with careful microsurgical technique.

The reported results of digital periarterial sympathectomy have generally been good. Egloff et al. (42) showed improvement in 13 patients who had Raynaud’s disease. In another series (43), 90% of patients had complete recovery, with ulcers healing in 3 weeks. Moreover, PVR and radionuclide scans showed marked increase in digital blood flow. Digital temperatures also improved by as much as 6°C within 2 days of operation. Carrying out digital periarterial sympathectomies for ischemic digital pain and ulcers in Raynaud’s disease, CREST syndrome, and ulnar artery thrombosis, other authors (44) reported relief of pain at 2 weeks and healing of ulcers at 3 months. Palmar periarterial sympathectomy or sympathectomy of radial and ulnar arteries at the wrist are safer options than operation on the fingers in patients with scleroderma or CREST syndrome.

However, Kaarela et al. (45) have not been able to demonstrate distal adrenergic denervation after periarterial sympathectomy in monkeys. They suggest that the positive effects of the operation are due to loss of adventitial support of the vasospastic arteries rather than adrenergic denervation.

Acrocyanosis is coldness and cyanosis of the hands and feet; it is not limited to the digits but involves the entire hand and foot. It is bilateral, symmetric, persistent, and worse in cold weather and does not entirely disappear with warmth. The pathology is based on arteriolar vasoconstriction with dilatation of the subpapillary venous plexus of the skin. Blood flows sluggishly in capillaries and veins. The distribution of acrocyanosis between men and women is approximately equal. It most often makes its appearance during the second or third decade of life. It improves with pregnancy and with advancing age. No treatment is generally required.

Erythromelalgia is a vasospastic disease that is characterized by episodes of erythema and a painful burning sensation in the area of the extremity involved. The skin becomes warm. The attack is initiated by direct or indirect heating of the extremity in which the skin is warmed above the critical point (usually from 32° to 36°C). There is a marked vasodilatation and a burning distress. Increasing the venous pressure with a tourniquet or through placement of the limb in a dependent position may produce symptoms. The burning pain and erythema may persist for many hours and are worse during the summer.

Primary erythromelalgia is of unknown etiology. Secondary erythromelalgia may exist with hypertension, polycythemia vera, gout, or heavy metal poisoning. Upper and lower extremities may be involved. Lower extremity involvement is more common in the primary form. In secondary erythromelalgia, only one extremity is usually affected.

Treatment of the primary form consists of nonspecific avoidance of warm environments. Elevation of the extremity and application of ice packs usually relieve the acute episode. Epinephrine may give dramatic relief. Acetylsalicylic acid can relieve some of the burning pain. Diagnosis of the underlying disease and its appropriate treatment is recommended in the secondary form of the disease. Erythromelalgia should be easily differentiated from a similar clinical appearance...
that is seen in some cases of arterial sclerosis and peripheral neuritis. 

Livedo reticularis is a peculiar response on the part of the small arteries that reach the skin that produces a reticulated or fishnet pattern of cyanosis that surrounds a paler central core. The cyanosis is caused by peripheral diluted venules that drain the capillary network that is supplied by the small arteries. It is distributed throughout the entire extremity, usually bilaterally, and, occasionally, on the trunk.

In its primary form, livedo reticularis is present from childhood and is completely asymptomatic. Secondly, it is seen after prolonged application of heat; after the administration of cortisone; in flaccid, paralytic limbs; and in limbs that have been immobilized in casts for a long time. Secondary livedo reticularis may suggest one of a large number of underlying diseases, which should be investigated, most of which were mentioned previously in the discussion of Raynaud’s phenomenon.

**COMPRESSIVE VASCULAR DISORDERS**

Thoracic outlet syndrome (TOS) is a phrase that is intended to replace a multiplicity of terms, such as cervical rib, prominent C-7 transverse process, costoclavicular, scalenus anticus, first rib, shoulder girdle, hyperabduction, pectoralis minor, and cervicobrahial neurovascular compression syndromes, among others. TOS results from compression of the brachial plexus and one or more arteries and veins, thereby producing varying symptoms of pain, numbness, and upper extremity ischemia.

The three most likely anatomic areas for compression of the neurovascular structures are the scalene triangle, the costoclavicular space, and the pectoralis minor space. The scalene triangle is bordered by the scalenus medius, the anterior muscles, and the first rib, into which it inserts. The brachial plexus and the second portion of the subclavian artery course over this rib and through the triangle. The subclavian vein is superficial to the scalenus anterior muscle. The costoclavicular space is that point of potential compression at which the clavicle, with its undersurface covered with a varying amount of subclavian musculature, passes over the first rib. This space narrows with clavicular depression or hyperabduction (as in inspiration). The pectoralis minor space is the least common point of compression. The pectoralis minor muscle and the coracoid process, where it inserts, may provide an unyielding fulcrum of compression, when the extremity is abducted (46).

Predisposing factors to thoracic outlet compression include (a) previous clavicular fracture with malunion, nonunion, or hypertrophic callus formation; (b) an anatomically long cervical spine that results in increased height for the second portion of the subclavian artery as it exits from the thorax; (c) neck muscle hypertrophy (47); (d) anatomic variations in the scalenus muscles and neurovascular structures; (e) a wide insertion of the scalenus anterior onto the first rib; (f) old cervical hypertension injury with trauma to and subsequent shortening and fibrosis of the scalenus musculature; (g) fractures of the first rib; (h) a hypertrophied subclavius muscle; (i) poor posture; (j) the aging process or debilitating diseases that decrease tissue elasticity; and (k) space-occupying tumors.

Symptoms are vague but consist of varying degrees of numbness or paresthesias in the hand and forearm. Frequent descriptions of symptoms that are given by patients are cramping, heaviness, or aching sensations in the forearm that are associated with pectoralis minor compression. The exaggerated military brace maneuver, with the shoulders drawn backward and downward, reproduces or aggravates symptoms at the costoclavicular level. When the test is positive, the patient’s hand is on the lap, (c) aneurysm that is signified by prominent supraclavicular pulsation or mass, (d) exclusion of cardiac sources of peripheral embolization, and (e) acute arterial insufficiency (47).

**Treatment**
TOS that is secondary to vascular compression is rare, as it is found in only 1% to 2% of all TOS patients. Because most occurrences of TOS are neurogenic in origin, the treatment modalities that are discussed in this section center on nerve decompression. The physician should follow TOS patients with repeated examination to reaffirm the nature of the complaint and the magnitude of the disability. Conservative measures benefit mild to moderate compression syndromes. In these cases, the patient may need to change occupations, as well as posture.

Patients with disabling symptoms that are refractory to conservative treatment require surgical correction, which commonly consists of resection of the first rib. Before surgery psychological consultation is requested to evaluate the patient's pain threshold. Radiographs are always taken. Magnetic resonance imaging becomes increasingly useful in preoperative evaluation, as experience is gained in its use. When scalenus anterior block with a long-acting anesthetic solution (bupivacaine) improves pain symptoms, surgical thoracic outlet decompression may be indicated. Scalenal blocks that use anesthetic solution and a steroid, however, may provide a degree of permanent relief to the patient with old trauma that resulted in fibrotic contracted muscle.

Surgical resection of the first rib is definitely preferred (46) over the cosmetically undesirable and less effective resection of a portion of the clavicle, unless a clavicle malunion has contributed directly to the symptoms of TOS; after clavicle malunion, corrective clavicular osteotomy is the treatment of choice. Before surgery psychological consultation is requested to evaluate the patient's pain threshold. Radiographs are always taken. Magnetic resonance imaging becomes increasingly useful in preoperative evaluation, as experience is gained in its use. When scalenus anterior block with a long-acting anesthetic solution (bupivacaine) improves pain symptoms, surgical thoracic outlet decompression may be indicated. Scalenal blocks that use anesthetic solution and a steroid, however, may provide a degree of permanent relief to the patient with old trauma that resulted in fibrotic contracted muscle.

As many as one-third of patients with TOS continue to have significant symptoms after rib resection. A certain percentage of these patients have abnormal scalene muscles and fibrous bands that compress the brachial plexus. Patients with conditions that are refractory to rib resection may be considered for scalene muscle and fibrous band decompression, if the symptoms warrant this treatment. Unfortunately, preexplorative differentiation of patients who may respond to first rib resection or to scalene muscle fibrous band decompression is difficult. Positioning of the cervical spine to discover areas of maximum tenderness may help to localize the compression site under some circumstances.

**CONGENITAL VASCULAR MALFORMATIONS**

In the past, a lot of confusion arose in the classification of congenital vascular anomalies. The epochal work of Mulliken and Glowacki (51) in 1975 has helped to clarify our understanding of these lesions. Their simplified classification has focused on endothelial characteristics on the basis of cell kinetics. They classified congenital vascular anomalies into two types: hemangiomas and malformations. Hemangiomas grow by cellular proliferation and tend to involute, whereas vascular malformations usually demonstrate normal endothelial growth characteristics throughout their natural history.

The authors further subdivided vascular malformations into capillary, venous, arterial, and lymphatic types (Table 1).

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**FIGURE 24.** Hemangioma of the right ring finger.

**Hemangioma**

Hemangioma usually seems to commence within the first few weeks of life. Rarely is a hemangioma fully formed at birth. From a humble beginning as an innocuous macular red spot, these lesions grow dramatically in the first year of life, only to become quiescent by 5 years of age (Fig. 24).

The color of the lesion depends on its location in relation to the skin. Lesions in the superficial dermis are called
strawberry hemangioma. Extremely superficial lesions may ulcerate. This occurs in 30% of upper limb hemangiomas (52). In finger suckers, the hemangioma may present as a chronic paronychial infection.

Within the first year, the lesion starts to involute, beginning with the formation of gray “herald spots” and followed by softening and lightening. Occasionally, the remaining fibrofatty tissue requires excision.

**Malformations**

Vascular malformations grow with the child. They are present at birth, but some may not be apparent. Malformations contain one or more vascular elements and may be classified into low-flow (capillary, lymphatic, and venous) or high-flow (arterial or fistula) lesions.

**Capillary**

Port wine “stain” is an example of a capillary malformation of the dermal vasculature. They can be diffuse or localized and usually appear pink in children. The treatment of these lesions is usually conservative and consists of the judicious use of cosmetics to mask the blemishes. Laser therapy may help in adults.

**Venous**

Venous malformations are the most common vascular malformations in the upper extremity (53). The spectrum of involvement varies from barely noticeable lesions to painful subcutaneous thrombosis and skin ulceration to engorgement on the assumption of a dependent position.

Treatment should be mainly conservative and consists of the use of elevation and compression garments. If surgical treatment is considered, extensive resection may be required. Careful microsurgical dissection should be carried out, with precautions taken to avoid hematoma. Staged surgical excision may be necessary in certain anatomic regions.

**Lymphatic**

Lymphatic lesions can range from small lymph-filled spaces to large cavities in the arm. The skin that overlies larger lesions is often altered in texture and thickness and may exhibit warty appearance. Although compression garments may be helpful in smaller lesions in the arm, larger malformations require staged surgical debulking. Complications of total excision and grafting include massive, and often unstable, scars, contracture, and ulceration. Lymphatico-venous anastomoses are being used in certain centers.

**Arterial**

Arterial malformations present infrequently in the upper extremity (53). Warm skin, hyperhidrosis, pulsating masses and thrills, or bruit constitute the usual presenting symptoms. The complications of extensive lesions include congestive heart failure and consumption coagulopathy. In the upper extremity, fingertip ischemic changes may occur owing to steal syndrome through fistulous tracts. Although noninvasive vascular studies determine relative flow distal to the lesion, angiography is the most informative preoperative investigation.

Pain and swelling are common presenting features. Pain occurs owing to venous engorgement (54), nerve entrapment (55), and spontaneous thrombosis (54, 55 and 56).

Initial treatment is conservative and consists of the use of pressure garments. With growth of the child, fistulous tracts tend to grow, thus giving rise to increasing pain and ischemia that is due to the increasing steal mechanism. Smaller lesions can be satisfactorily treated surgically. Proximal ligation is often unsuccessful in redirecting the flow and can actually worsen the condition. Partial staged excision with revascularization and skin flaps has been shown to give the most favorable results in selected cases. Many authors have reported high rates of recurrence and complications after surgical excision of malformation (57-58). Hill et al. (57) reported a recurrence rate of 47%. Recurrences were more frequent in the diffuse type, and when excision was considered incomplete. These authors recommend surgery only for disabling symptoms that are not controlled by conservative treatment. Although selective embolizations have been used in certain centers, the results are unpredictable and can be complicated by distal ischemia (52).

**OCCUPATIONAL VASCULAR DISORDERS**

Vibration white finger (VWF) is a form of secondary Raynaud’s phenomenon that is induced by transmission of vibration energy from handheld vibratory tools to the hand and forearm (60). Pneumatic tools were introduced in France in 1839 and were widely used after 1880. Electric and gasoline motors were used in vibration tools in the 1940s. Motored chain saws were first manufactured in 1958. In 1911, Loriga (61) first reported that Italian quarrymen who used vibratory tools developed symptoms similar to those of Raynaud’s phenomenon. In 1939, Leys (62) reported the first case of combined Raynaud’s phenomenon and diffuse scleroderma and the use of vibratory tools. In 1918, Calle et al. (63) investigated the limestone cutters of Bedford, Indiana. Since 1946, it has been a prescribed disease in Denmark (64). VWF became a prescribed occupational disorder in Japan in 1966 and in the United Kingdom in 1985 (65).

Raynaud’s phenomenon that occurs owing to exposure to vibration is now called hand-arm vibration syndrome. The acceleration range of vibration in hand-arm vibration syndrome is much higher than it is in whole body vibration. This has been measured from 1g to as much as 2,400g, with a frequency range from 6 to 1,250 Hz.

Manifestations of prolonged vibration in the upper limb include (a) Raynaud’s phenomenon, (b) peripheral neuropaths, (c) muscle injury and damage to joints, and (d) bone changes. The effects depend on the physical characteristics of vibration, such as the frequency spectrum, magnitude, and direction of vibration. Other factors that influence the development of Raynaud’s phenomenon are the type of tool, the posture of the hands, the daily duration of exposure, the temporal exposure pattern, the duration of cumulative pattern, and the environmental conditions, including temperature and noise levels (61).

Pyykko (66) calculated the latency to be 5,600 plus 2,500 hours of exposure for development of VWF. Miya-shita et al.
(67), studying chain saw workers, found that less than 2,000 hours of cumulative exposure to vibration led to symptoms of tingling, numbness, and pain; between 2,000 and 5,000 hours of exposure led to peripheral nerve and circulatory disturbances; and between 5,000 and 8,000 hours of exposure led to functional changes. When the cumulative exposure was more than 8,000 hours, approximately one-half of the operators experienced severe functional or organic changes due to vibration. In studying platers, Nilsson et al. (68) found that 8% of the workers had VWF after 4 years of exposure to vibration, 84% had VWF after 5 to 9 years, and 94% had VWF after 10 to 19 years. According to the authors, each year of vibration exposure raises the odds ratio for VWF by 11%. In rock drillers of British Columbia, Brubaker et al. (69) found a prevalence rate of 25% after 1 to 5 years of vibration exposure. This increased to 80% after 16 years of exposure.

VASCULAR TUMORS

Benign Tumors

Glomus tumors comprise 1% to 4.5% of all hand tumors and are more common in adults. They probably arise from a normal structure, the myoarterial glomus that is present in the stratum reticulare of the dermis, which has a role in temperature regulation. Whereas the normal glomus is less than 1 mm in diameter, the tumor may be as large as 1 cm. The distribution of glomus tumors follows the distribution of the precursor glomus, with as much as 75% present in the hand, especially in the distal segment of the digits. A subungual location is common (Fig. 25). Detailed radiographs may demonstrate underlying bony erosion. A small area of bluish discoloration that is tender to pressure may be seen deep in the nail.

The typical triad is pain that lancinates with proximal radiation, sensitivity to temperature change, and exquisite tenderness. Probing the digit with pressure from the head of a pin may be used to localize the site of the tumor. When the pinhead comes to rest directly over the tumor, the patient experiences exquisite tenderness.

Treatment of glomus tumors is excision. Recurrence rates as high as 18% to 25% have been reported (60). These rates can be reduced by searching for multiple tumors and using magnification (microscope) to ensure total removal of the glomus (Fig. 15).

Hemangiopericytoma

Hemangiopericytoma is a rare vascular tumor that is derived from cells that surround capillaries and postcapillary venules or pericytes (52). Ovoid- or spindle-shaped tumor cells proliferate around endothelial-lined vascular channels. These tumors are difficult to diagnose histologically. There are three types of biologic activity of the tumor: benign, borderline malignant, and malignant (70). This lesion has a characteristic appearance on magnetic resonance imaging and contrast angiography (54). Management of the lesion depends on grade and consists of a combination of surgery, chemotherapy, and radiation.

Dermal Angiomyoma

Dermal angiomyomata are actually vascular leiomyomas and, in the extremity, arise from the smooth muscle of arterial walls. They present as painless, slow-growing masses and are located in the deep dermis or subcutaneous tissue. At surgery, they appear as well-encapsulated, white, round, or oval tumors that are 1 to 3 cm in size. Treatment is excisional biopsy. Recurrence should alert one to the possibility of a leiomyosarcoma.

Vascular Leiomyomas

Vascular leiomyomas or angioleiomyomas are benign tumors of smooth muscle origin that arise from the muscular layer of the vessel wall (63). Histologically, the tumor contains smooth muscle bundles with surrounding vascular channels. Calle et al. (63) recommended a 2- to 3-mm margin of resection. No recurrence has been reported after excision of the tumor.

Malignant Tumors

Malignant vascular tumors of the upper extremity have been reported but are infrequently seen. Hemangiosarcoma, hemangioendotheliosarcoma, and hemangiopericytoma, as well as Kaposi’s sarcoma, do occur (62), however, and...
should be treated with the same principles that govern cancer surgery in other locations. Tumor growth that is more rapid than the normally slow progression of a vascular lesion, an increase in pain, signs of local ischemia, and even infections complications should alert one to the possibility of malignant change.

### Angiosarcomas

Angiosarcomas are characterized by rapid growth. They are widely metastatic. Usually, a high-level amputation is recommended. However, the prognosis of these lesions is poor (71).

### Kaposi’s Sarcoma

Kaposi’s sarcoma is seen in two groups of patients: (a) as an acquired immune deficiency syndrome complex, where it starts in the head or neck region, and (b) in elderly men of Jewish or Mediterranean descent (71). It presents as multiple small purplish lesions and is responsive to surgical or radiation therapy. A condition that is called angiolymphoid hyperplasia with eosinophilia can present a pathologic picture that is similar to Kaposi’s sarcoma (8). This lesion is considered a benign inflammatory lesion and should not be confused with Kaposi’s sarcoma.

### Lymphangiosarcomas

Lymphangiosarcomas are seen in long-standing postmastectomy lymphadenopathy. They are present initially as bluish lesions that ulcerate and undergo rapid metastasis. Early radical amputation becomes necessary.

### VENOUS OCCLUSIVE DISEASE

Deep vein thrombosis of the upper limb, 2% of total deep vein thrombosis in the body (72). Thrombosis of the veins of the forearm occurs frequently. Such veins may be perceived during diagnosis as palpable cords. This condition may take the form of thrombosis alone, phlebothrombosis, or, in association with surrounding inflammation, thrombophlebitis. Phlebothrombosis is occasionally seen in the dehydrated individual, whereas thrombophlebitis is seen frequently as a complication of intravenous therapy. In cases of thrombophlebitis, there is surrounding pain, tenderness, and erythema as well. In the digits, the presence of a tender palpable nodule or cord sometimes follows an episode of trauma.

Treatment is generally symptomatic and consists of removal of any offending intravenous catheter, application of warm packs, and elevation of the extremity. For persistent tenderness that does not respond to such therapy, or in patients with supplicative thrombophlebitis, excision of the involved vein is indicated.

Thrombosis of the axillary or subclavian veins or the superior vena cava may manifest acutely in the upper extremity with edema, discoloration, from nearly normal to cyanosis; dilatation of the venous pattern; and pain. Spontaneous thrombosis of the subclavian vein may occur after exertion, commonly in the dominant extremity, in the middle-aged obese female with pendulous breasts, and in patients who have poor posture or loss of muscle tone. Occasionally, however, this is seen in the young muscular weight lifter. Unusual activity, such as carrying a heavy suitcase, often precedes the onset of symptoms. Thrombosis is usually due to compression of the subclavian vein in the thoracic outlet as it passes between the clavicle and first rib.

Other causes of large vein thrombosis include underlying systemic diseases, lymphatic and venous involvement with neoplastic disease, such as Pancoast’s tumor, and thrombophlebitis of varying etiology from indwelling catheters or injection of irritating drugs. Correction of these precipitating factors is necessary to prevent chronic thrombophlebitis.

The diagnosis of thrombosis can be confirmed by venography. Noninvasive venous studies represent an additional helpful screening tool. If a diagnosis of acute axillary or subclavian vein thrombosis is established within 12 hours of the onset of symptoms, the surgeon must seriously consider prompt venous thrombectomy to prevent late sequelae. Associated areas of venous compression or constriction are corrected at the time of thrombectomy.

Patients who have mild symptoms or who are diagnosed late are treated conservatively, that is, with elevation of the extremity, warm packs, and intravenous heparin, and sympathetic blocks, if there is an element of associated vasospasm. Concurrent use of sympathetic block and heparin should be avoided. If this is not possible, a long-acting stellate block should precede heparin administration. If a stellate block is used initially to relieve vasospasm, it should be followed by long-term elevation of the extremity and intravenous heparin therapy.

Additionally, intravenous heparin is administered to those patients who have undergone venous thrombectomy. If the thrombus should recur, the patient improves through administration of heparin, which appears to be associated with the dilatation of venous collateral channels. The role of the thrombolysins, streptokinase and urokinase, in the treatment of upper extremity venous thrombosis is still controversial. See the section Thrombosis for a discussion of their relative effectiveness in lysing a clot and restoring patency.

Recurrent vein thrombosis that initially is treated conservatively should be studied with venography. This often reveals a point of vein constriction near the thoracic outlet area that necessitates surgical correction, as was described in the previous discussion. One must also be aware that patients with recurrent thrombophlebitis may have underlying malignancy.

Late sequelae of large vein thrombosis are persistent brawny edema, coldness, pain, and limitation of motion of the extremity. These symptoms can, to some extent, be alleviated by elevation, compression garments, sympathetic blocks, and surgical release of areas of venous compression.

The patient who presents with superior vena caval obstruction that is secondary to a malignant process represents one of the few indications for immediate radiation therapy in the area of the tumor. In addition, the patient is treated with the previously described conservative measures. Rarely is surgical bypass necessary.

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