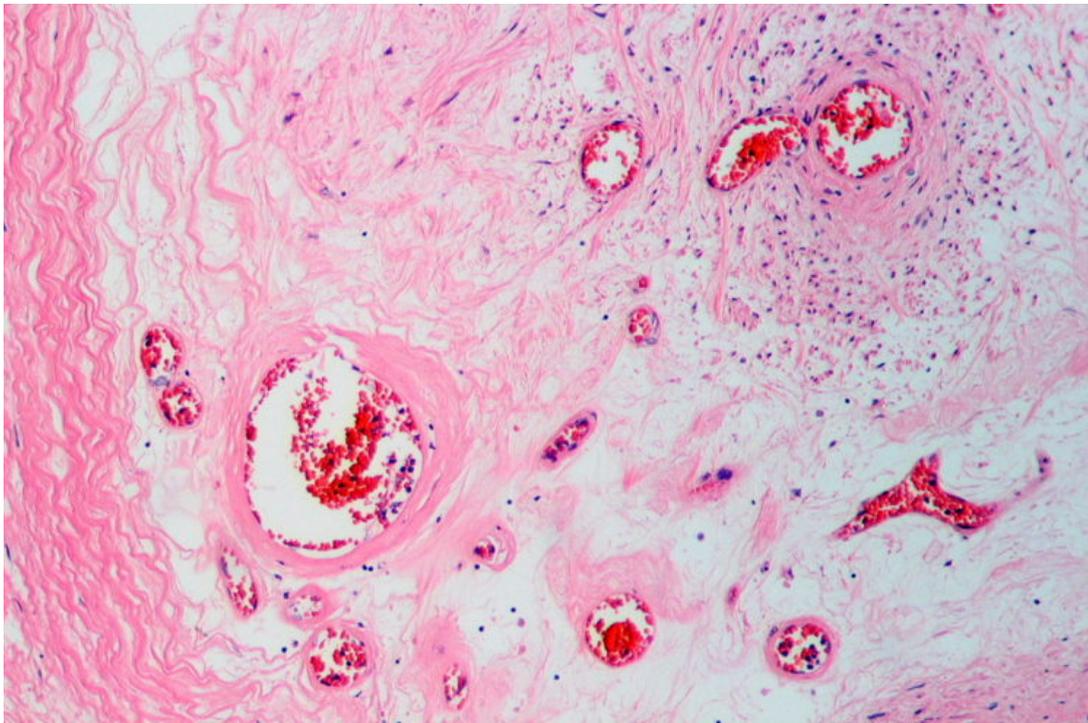


Overview of Vascular Diseases

[See online here](#)

Vascular diseases are any abnormal conditions that affect the circulatory system and are responsible for more mortality and morbidity than any other category of human disease. The spectrum of pathology encompasses congenital and acquired disorders that are arterial, venous, capillary or lymphatic in origin. Vascular disease can affect virtually any vessel in the body and present with different symptoms in different regions of the body. The most commonly involved areas include the heart, head, neck, and upper and lower extremities. Less commonly, vessels supplying the bowel may be involved leading to bowel infarction.



Definition of Terms

- **Thrombosis** - the formation or presence of a blood clot (thrombus) in a blood vessel
- **Stenosis** - the narrowing/tightening of the lumen of a blood vessel
- **Occlusion** - obstruction/blockage of a blood vessel
- **Dissection** - a tear within the wall of a blood vessel, which allows blood to separate the wall layers
- **Necrosis** - the death of tissue
- **Ischemia** - reduction/restriction in blood supply

Arterial Diseases

Raynaud's Syndrome

This refers to **peripheral digital ischemia** due to **vasospasms** precipitated by stress or cold. Fingers or toes ache and change color from pale to blue, to red.

It may be idiopathic ([Raynaud's Disease](#)/Primary Raynaud Phenomenon), characterized by the occurrence of the vasospasm alone, with no association with another illness. There may be an underlying cause (Secondary Raynaud Phenomenon) usually an **autoimmune disease**, such as **systemic sclerosis** (90% of individuals with scleroderma), **systemic lupus erythematosus (SLE)**, [rheumatoid arthritis](#) and **dermatomyositis/polymyositis**.

Buerger's Disease



Image: "55-year-old female with advanced Buerger's disease. Two toes amputated due to gangrene. Both legs scheduled for amputation." by Onthelist - Own work, License: [CC BY-SA 4.0](#)

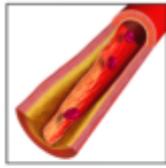
This is also referred to as **thromboangiitis obliterans (TAO)**. It is a **smoking-related inflammatory vasculopathy**, characterized by an inflammatory endarteritis that causes a pro-thrombotic state and subsequent vaso-occlusive phenomena. It characteristically affects medium-sized arteries, as well as veins of the upper and lower extremities (which thrombose, causing gangrene).

Patients often present with **moderate-to-severe claudication** that can quickly progress to **critical limb ischemia** featuring rest pain or tissue loss. Abstinence from tobacco is the only measure known to prevent disease progression.

Carotid artery stenosis

This usually occurs as a result of **atherosclerotic disease** of the [carotid artery](#), accounting for 20% of **strokes and transient ischemic attacks**. Symptomatic patients with ipsilateral stenosis $\geq 70\%$ should have carotid endarterectomy.

Peripheral artery disease



[Image](#): "Peripheral artery disease" by Blausen.com staff. "Blausen gallery 2014". Wikiversity Journal of Medicine. DOI:10.15347/wjm/2014.010. ISSN 20018762. - Own work, License: [CC BY 3.0](#)

Claudication, which is defined as reproducible ischemic muscle pain, is one of the most common manifestations of **peripheral arterial occlusive disease (PAOD)** caused by atherosclerosis.

Claudication occurs during physical activity and is relieved after a short rest. **Ankle-brachial index (ABI)** is a useful non-invasive way of establishing the presence of PAOD and is calculated as the ratio of systolic blood pressure at the ankle to that in the arm (normal range, 0.9-1.1; PAOD, <0.9). **Angiography** is the criterion standard arterial imaging study for the diagnosis of PAOD. Management may be:

- Conservative: regular exercise, tobacco cessation, control of lipid profile, [diabetes](#), and hypertension. Anti-platelets agents have a role (usually aspirin).
- Surgical: open bypass surgery or endovascular therapy (e.g. stents, balloons, or atherectomy devices)

Inflammatory Diseases

An inflammatory disorder of the [blood vessel](#) is referred to as a **vasculitis**. It presents with **systemic manifestations** (including fever, malaise, myalgias, and arthralgias) and **organ dysfunction** that depends on the pattern of vascular involvement. It should, therefore, be considered in any unidentified multisystem disorder.

Most vasculitis responds to **steroid therapy**. Vasculitis can result from **infections**, but, more commonly, has an **immunologic basis** such as immune complex deposition, anti-neutrophil antibodies (ANCA), or anti-endothelial cell antibodies. Vasculitis may also be

secondary to other diseases such as **SLE, rheumatoid arthritis, hepatitis B & C,** and **HIV.**

Different forms of vasculitis tend to specifically affect vessels of a particular caliber and location. They are categorized as follows, depending on the size of the blood vessel affected:

1. **Large vessel vasculitis:** giant cell arteritis and Takayasu arteritis
2. **Medium vessel vasculitis:** Polyarteritis Nodosa and Kawasaki disease.
3. **Small vessel vasculitis:**
 - ANCA positive: microscopic polyangiitis, Wegener's Granulomatosis, Churg-Strauss Syndrome
 - ANCA negative: Henoch-Schonlein Purpura, Goodpasture's Syndrome, cryoglobulinemia

Giant cell arteritis (GCA)

This is also known as **cranial or temporal arteritis.** It is associated with **polymyalgia rheumatic** in 50% of individuals. Common signs and symptoms of GCA include visual disturbances, headache, jaw claudication, neck pain, and scalp tenderness.

It should always be considered in the differential diagnosis in patients 50 years of age or older with an **elevated erythrocyte sedimentation rate.** If suspected, **steroid therapy (prednisolone)** should be started immediately as failure to do so **risk irreversible bilateral visual loss.** **Temporal artery biopsy** is the criterion standard for the diagnosis of this granulomatous vasculitis.

Takayasu arteritis

This is also known as an **aortic arch syndrome** or **pulseless disease.** It is a rare, systemic, inflammatory large-vessel vasculitis of unknown etiology that most commonly affects women of childbearing age (commonly 20-40 years old). It usually affects the aorta and its major branches, causing **stenosis** and **thrombosis.** Acute inflammation causes **dilatation** and **aneurysms.** Its incidence is higher in Asian countries, such as Japan, and rarer elsewhere.

Polyarteritis Nodosa (PAN)

This is a systemic vasculitis characterized by **necrotizing inflammatory lesions** that affect medium-sized arteries resulting in **microaneurysm formation,** an **aneurysmal rupture with hemorrhage, thrombosis,** and, consequently, **organ ischemia or infarction.** Like other vasculitides, PAN is a multi-system disorder with a myriad of manifestations, although it commonly affects the skin, joints, the gut, and the kidneys. PAN may be associated with **hepatitis B.**

Kawasaki disease

This is an **acute febrile vasculitic syndrome** of early childhood (median age of 10 months), characterized by the inflammation of medium to small-sized arteries causing aneurysms to form. Prognosis is good with treatment (**aspirin or intravenous immunoglobulin**), although, in a small percentage of patients, it can lead to death resulting from **coronary artery aneurysms (CAA).** Coronary arteritis can cause aneurysms that rupture or thrombose, resulting in **myocardial infarction.**

Microscopic polyangiitis

This is also known as **hypersensitivity vasculitis** or **leukocytoclastic vasculitis**. It is a necrotizing vasculitis affecting small-sized vessels. **Necrotizing glomerulonephritis** (seen in 90% of patients) and **pulmonary capillaritis** are particularly common.

Wegener's granulomatosis

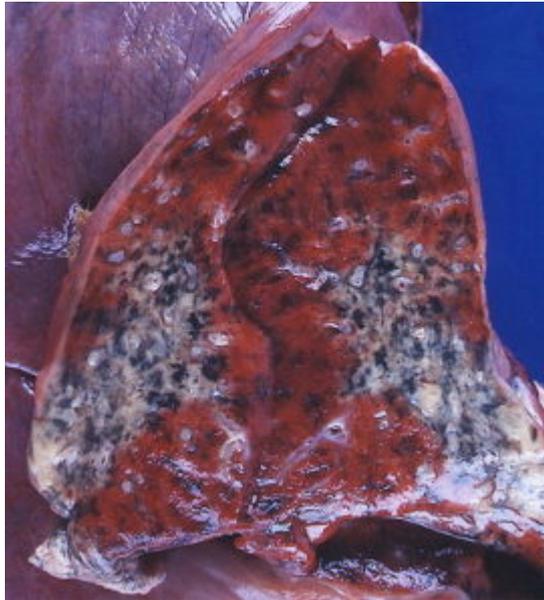


Image: "Wegener's granulomatosis" by Yale Rosen from USA, uploaded by CFCF, License: [CC BY-SA 2.0](https://creativecommons.org/licenses/by-sa/2.0/)

This is a **necrotizing vasculitis** characterized by a specific triad of findings:

- **Granulomas** of the [lung](#) and/or upper respiratory tract (ear, nose, sinuses, and throat).
- **Vasculitis** of small to medium-sized vessels, most prominently in the lungs and upper [respiratory tract](#)
- **Glomerulonephritis**

Clinically, it **resembles Polyarteritis Nodosa** with the additional feature of **respiratory involvement**.

Churg-Strauss Syndrome

This is also known as **allergic granulomatosis** and **angiitis**. It is a small vessel vasculitis classically associated with [asthma](#), [eosinophilia](#), and **granulomas**.

Henoch-Schonlein Purpura (HSP)

This is a systemic variant of **IgA nephropathy (Berger's Disease)**, causing a small vessel vasculitis. It usually presents with a **purpuric rash** on extensor surfaces (typically on the legs) and [glomerulonephritis](#). Diagnosis is usually clinical, but can be confirmed with **positive immunofluorescence for IgA and C3 in the skin or renal biopsy**.

Goodpasture's Syndrome

This is also known as **anti-glomerular basement membrane (GBM) disease**, and, as the name suggests, it is caused by the development of autoantibodies to type IV collagen, an essential component of the GBM. This autoimmune disorder is characterized by autoantibodies directed against the glomerular and the alveolar basement membrane, hence patients present clinically with manifestations of both **pulmonary and renal disease**, which may include **hemoptysis** and **hematuria**.

Cryoglobulinaemia

This is characterized by the presence of **cryoglobulins in the serum**. This may result in a clinical syndrome of **systemic inflammation** (most commonly affecting the [kidneys](#) and skin) caused by cryoglobulin-containing immune complexes. It may be associated with **hepatitis C**.

Diseases of the Aorta

Aortic dissection



[Image](#): "CT reconstruction of aortic dissection" by Dr. Lars Grenacher (www.grenacher.de), uploaded by J. Heuser - scan courtesy of Dr. Lars Grenacher (www.grenacher.de), License: [CC BY-SA 3.0](#)

[Aortic dissection](#) is defined as the separation of the layers within the aortic wall. Blood splits the aortic media with **sudden tearing chest pain** (with/without radiation to the back between the scapulae).

Aortic dissections occur mainly in two age groups: (1) men aged 40 to 60 years with antecedent hypertension (more than 90% of cases); and (2) younger patients with connective tissue abnormalities that affect the aorta (e.g. **Marfan Syndrome**). Dissections can also be iatrogenic (e.g. complicating arterial cannulation), and rarely, for unknown reasons, may occur in **pregnant women**. Aortic dissections are generally classified into two types:

- **Proximal lesions:** type A dissections, involving the ascending aorta, with or without the involvement of the descending aorta (DeBakey type I or II, respectively)
- **Distal lesions:** type B dissections, usually beginning beyond the subclavian artery (DeBakey type III).

All patients with type A dissections should be considered for **surgery and urgent cardiothoracic advice** sought. Type B can be managed conservatively.

Aneurysms

Aortic aneurysms are congenital or acquired dilatations of the heart or blood vessels. Aneurysms of the aorta are **fusiform**, which are **circumferential dilations** that can be up to 20 cm in diameter.

The two most important causes of aortic aneurysms are **atherosclerosis** and **hypertension**. Other conditions that weaken vessel walls and lead to aneurysms include **trauma, vasculitis, congenital defects, and infections** (the so-called **mycotic aneurysms**). Complications are related to **rupture, thrombosis, and embolization**.

Renal artery stenosis

This is the major cause of renovascular hypertension. The patient presents with **refractory hypertension** (i.e. poor control of hypertension, despite treatment with three or more antihypertensive agents). It tends to occur in older individuals, with **atherosclerosis** being, by far, the most common etiology of renal artery stenosis.

Venous Diseases

Pulmonary embolism

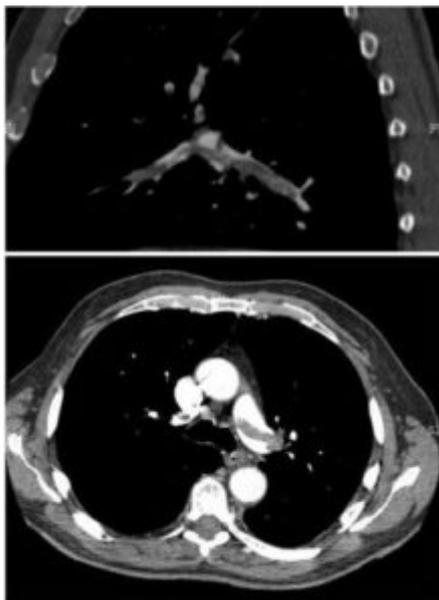


Image: "CT pulmonary angiography images confirming the presence of a saddle embolus and substantial thrombus burden in the lobar branches of both main pulmonary arteries." by Aung Myat and Arif Ahsan - Percutaneous mechanical thrombectomy for the treatment of

Pulmonary embolism occurs when clots break off and pass through the veins and the right side of the heart before lodging in the pulmonary circulation. They usually arise from thrombi originating in the deep venous system of the lower extremities; however, other rarer origins include thrombi from the pelvic, renal, upper extremity veins, the right heart chambers (post-MI); septic emboli (**right-sided endocarditis**); fat, air, amniotic fluid embolism; neoplastic cells and parasites.

The classic presentation of pulmonary embolism is an abrupt onset of **pleuritic chest pain, shortness of breath** and **hypoxia**.

Risk factors include recent surgery (especially abdominal/pelvic or hip/knee replacement), **thrombophilia** (e.g. antiphospholipid syndrome), leg fracture, prolonged bed rest/reduced mobility, malignancy, pregnancy/postpartum, hormone replacement therapy, and a previous pulmonary embolism. Management of this involves **anticoagulation** (e.g. using low-molecular-weight heparin) and **thrombolysis** (e.g. alteplase).

Deep venous thrombosis (DVT)

DVT is a manifestation of venous thromboembolism (VTE). Although most DVT is occult and resolves spontaneously without complication, lots of deaths result from DVT-associated massive pulmonary embolism (PE).

Risk factors include recent surgery (especially abdominal/pelvic or hip/knee replacement), trauma, thrombophilia (e.g. antiphospholipid syndrome), leg fracture, prolonged bed rest/reduced mobility, malignancy, pregnancy, obesity, hormone replacement therapy/synthetic estrogen, or a previous DVT.

Diagnosis is based on validated clinical prediction rules (e.g. the **Wells score**) and **D-dimer testing** (which is sensitive, but not specific for DVT as it is also increased in infections, pregnancy, malignancy, and post-op). A negative D-dimer result, combined with a low pretest clinical probability score is sufficient to exclude DVT. **Anticoagulation** is the mainstay of therapy. **Inferior vena caval filters** may be used in active bleeding, or when anticoagulation fails, to minimize the risk of **thromboembolism**.

Varicose veins

This refers to the long tortuous and dilated veins of the superficial venous system. The superficial veins of the upper and lower leg typically are involved, and the causes include:

- Primary causes (95%): unknown, **congenital valve absence** (very rare)
- Secondary causes (5%): **obstruction**: DVT, fetus, ovarian tumor; **valve destruction**: DVT; **arteriovenous malformation, constipation, overactive muscle pumps** (e.g. cyclists)

Pathology of varicose veins of the extremities is as follows: Valves prevent blood from passing from deep to superficial veins via perforator veins. If they become incompetent, there is **venous hypertension** and **dilatation of the superficial veins** occur. Risk factors include prolonged standing, obesity, pregnancy, family history, and the pill.

Varicosities (venous dilations) of other sites worth mentioning include esophageal varices which occur secondary to portal hypertension. They are important as they are

prone to ruptures that can lead to massive (even fatal) **upper gastrointestinal hemorrhage**.

Congenital Malformations

Hemangioma



Image: "Small hemangioma on the scalp of a two-year-old female." by Cbheumircanl - Own work, License: [Public Domain](#)

These are very common **benign tumors** composed of blood-filled vessels. They are usually present from birth and initially increase in size, but many eventually regress spontaneously.

Although these localized lesions are typically confined to the head and neck, they occasionally may be more extensive (**angiomatosis**) and can arise internally, with most of these internal lesions being found in the liver. Malignant transformation is rare. Histological/clinical variants include:

- **Capillary hemangiomas:** commonest; occur in the skin, subcutaneous tissues, and mucous membranes of the oral cavities and lips, as well as in the [liver](#), spleen, and kidneys.
- **Juvenile hemangiomas:** (so-called strawberry hemangiomas) of the newborn skin.
- **Pyogenic granulomas**

Cavernoma (cavernous hemangioma)

These are composed of **large, dilated vascular channels**. Compared to other hemangiomas, cavernous hemangiomas are more **infiltrative**, frequently involve **deeper structures** and do not spontaneously regress. Of note, cavernous hemangiomas constitute one component of **von Hippel-Lindau disease**, in which vascular lesions are commonly found in the cerebellum, brain stem, retina, [pancreas](#), and liver.

Malformations of the aorta

Congenital malformations of the aorta include **aortic valvular stenosis** or **complete-aortic valvular atresia**, **coarctation of the aorta**, **double aortic arch**, **right aortic arch**, and an **interrupted aortic arch**.

Worth mentioning is the **coarctation of the aorta** with an estimated incidence of 3.2/10,000 births. In this malformation, the aortic lumen below the origin of the left subclavian artery is significantly narrowed. It is distinguished into two types based on the level of constriction in relation to the entrance of the **ductus arteriosus** as **preductal** and **postductal**.

Diseases of the Lymphatic System

Elephantiasis



[Image](#): "Elephantiasis of legs" by Otis Historical Archives of "National Museum of Health & Medicine" (OTIS Archive 1) - <http://www.flickr.com/photos/medicalmuseum/3382983713/in/photostream>, License: [CC BY 2.0](#)

This refers to **leg lymphedema** resulting from **lymphatic filariasis**. The most frequent organisms causing this disease are *Wuchereria bancrofti* and *Brugia malayi*. These **filarial worms** block the flow of lymph, causing edematous arms, legs, and scrotum. The disease is transmitted by the bite of the infected *Anopheles* and *Culex* mosquitoes. Filariasis is diagnosed by **detection of microfilariae in blood**. It is treated with a **combination of diethylcarbamazine and albendazole**.

Cysts (cavernous lymphangiomas/cystic hygromas)

These benign lesions are composed of **massively dilated lymphatic spaces** lined by endothelial cells and are typically found in the neck or axilla of children. They can be very large, filling the axilla or producing gross deformities of the neck. Cavernous lymphangiomas of the neck are common in **Turner Syndrome**.

Review Questions

The correct answers can be found below the references.

1. **A 23-year-old man presents with fever, weight loss, malaise, abdominal pain, and myalgias. Workup reveals that the patient has Polyarteritis Nodosa. Which of the following is associated with this form of vasculitis?**

- A. Arsenic
- B. Chlamydia pneumonia
- C. Hepatitis B virus
- D. Human herpes-virus 8
- E. Human immunodeficiency virus

2. **A 3-year-old boy is brought by his parents to the emergency department because they are concerned that he has had a high fever for several days. On examination, the boy has conjunctival and oral erythema. He has palpable cervical lymphadenopathy and erythema of his palms and soles. What is a potentially life-threatening complication of this disorder?**

- A. Aneurysms of the abdominal aorta
- B. Aneurysms of the coronary arteries

- C. Dissection of the thoracic aorta
- D. Ischemia and gangrene of the extremities
- E. Rupture of a berry aneurysm.

3. A 32-year-old African American woman returns to her physician for a follow-up of hypertension that has been poorly controlled in spite of numerous antihypertensive medications. It is decided to evaluate the patient for possible “secondary” hypertension. Which of the following is a well-known cause of secondary hypertension?

- A. Ethnicity
- B. Obesity
- C. Renal artery stenosis
- D. Smoking
- E. Stress

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[Kawasaki Disease](#) via medscape.com

[Renal Artery Stenosis](#) via medscape.com

Correct answers: 1C, 2B, 3C

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