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Diseases of the Aorta

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The aorta is the conduit through which the blood ejected from the left ventricle is delivered to the systemic arterial bed. In adults, its diameter is approximately 3 cm at the origin, 2.5 cm in the descending portion in the thorax, and 1.8 to 2 cm in the abdomen. The aortic wall consists of a thin intima composed of endothelium, subendothelial connective tissue, and an internal elastic lamina; a thick tunica media composed of smooth-muscle cells and extracellular matrix; and an adventitia composed primarily of connective tissue enclosing the vasa vasorum and nervi vascularis. In addition to its conduit function, the viscoelastic and compliant properties of the aorta also subserve a buffering function. The aorta is distended during systole to enable a portion of the stroke volume to be stored, and it recoils during diastole so that blood continues to flow to the periphery. Because of its continuous exposure to high pulsatile pressure and shear stress, the aorta is particularly prone to injury and disease resulting from mechanical trauma (Table 231-1). The aorta is also more prone to rupture than any other vessel, especially with the development of aneurysmal dilatation, since its wall tension, as governed by Laplace's law (i.e., proportional to the product of pressure and radius), would be increased.

TABLE 231-1 Diseases of the Aorta: Etiology and Associated Factors

Aortic aneurysm
Atherosclerosis
Cystic medial necrosis
Tuberculosis
Syphilitic infection
Mycotic infection
Rheumatic aortitis

Trauma
Aortic dissection
Cystic medial necrosis
Systemic hypertension
Atherosclerosis
Takayasu's arteritis
Giant cell arteritis
Aortic occlusion
Atherosclerosis
Thromboembolism
Aortitis
Syphilitic aortitis
Rheumatic aortitis
Takayasu's arteritis
Giant cell arteritis

AORTIC ANEURYSM

An *aneurysm* is defined as a pathologic dilatation of a segment of a blood vessel. A *true aneurysm* involves all three layers of the vessel wall and is distinguished from a *pseudoaneurysm*, in which the intimal and medial layers are disrupted and the dilatation is

lined by adventitia only and sometimes by perivascular clot. Aneurysms may also be classified according to their gross appearance. A *fusiform aneurysm* affects the entire circumference of a segment of the vessel, resulting in a diffusely dilated lesion. In contrast, a *saccular aneurysm* involves only a portion of the circumference, resulting in an outpouching of the vessel wall. Aortic aneurysms are also classified according to location, i.e., abdominal versus thoracic. Aneurysms of the descending thoracic aorta are usually contiguous with infradiaphragmatic aneurysms and are referred to as *thoracoabdominal aortic aneurysms*.

ETIOLOGY

The most common pathologic condition associated with aortic aneurysm is *atherosclerosis*. It is controversial whether atherosclerosis itself actually causes aortic aneurysms or whether atherosclerosis develops as a secondary event in the dilated aorta. Causality is implied by studies that have shown that many patients with aortic aneurysms have coexisting risk factors for atherosclerosis (Chap. 224), particularly cigarette smoking, as well as atherosclerosis in other blood vessels. Seventy-five percent of atherosclerotic aneurysms are located in the distal abdominal aorta, below the renal arteries.

Cystic medial necrosis is the term used to describe the degeneration of collagen and elastic fibers in the tunica media of the aorta, as well as the loss of medial cells that are replaced by multiple clefts of mucoid material. Cystic medial necrosis characteristically affects the proximal aorta, results in circumferential weakness and dilatation, and leads to development of fusiform aneurysms involving the ascending aorta and the sinuses of Valsalva. This condition is particularly prevalent in patients with Marfan syndrome and Ehlers-Danlos syndrome type IV (Chap. 342) but also occurs in pregnant women, in patients with hypertension, and in those with valvular heart disease. Sometimes it appears as an isolated condition in patients without any other apparent disease. Familial clusterings of aortic aneurysms occur in 20% of patients, suggesting a hereditary basis of the disease. Mutations of the genes encoding fibrillin-1 and type III procollagen have been implicated in some cases. Linkage analysis has identified loci on chromosomes 5q13-14 and 11q23.3-q24 in several families, although the specific culprit genes have not been described. *Syphilis* (Chap. 153) is a relatively uncommon cause of aortic aneurysm. Syphilitic periaortitis and mesoaortitis damage elastic fibers, resulting in thickening and weakening of the aortic wall. Approximately 90% of syphilitic aneurysms are located in the ascending aorta or aortic arch. *Tuberculous aneurysms* (Chap. 150) typically affect the thoracic aorta and result from direct extension of infection from hilar lymph nodes or contiguous abscesses or from bacterial seeding. Loss of aortic wall elasticity results from granulomatous destruction of the medial layer. A *mycotic aneurysm* is a rare condition that develops as a result of staphylococcal, streptococcal, or salmonella infections of the aorta, usually at an atherosclerotic plaque. These aneurysms are usually saccular. Blood cultures are often positive and reveal the nature of the infecting agent.

Vasculitides associated with aortic aneurysm include Takayasu's arteritis and giant cell arteritis, which may cause aneurysms of the aortic arch and descending thoracic aorta. Spondyloarthropathies such as ankylosing spondylitis, rheumatoid arthritis, psoriatic arthritis, relapsing polychondritis and Reiter's syndrome are associated with dilatation of

the ascending aorta. Behçet's disease (Chap. 307) causes thoracic and abdominal aortic aneurysms. *Traumatic aneurysms* may develop after penetrating or nonpenetrating chest trauma and most commonly affect the descending thoracic aorta just beyond the site of insertion of the ligamentum arteriosum. *Congenital aortic aneurysms* may be primary or associated with anomalies such as a bicuspid aortic valve or aortic coarctation.

THORACIC AORTIC ANEURYSMS

The clinical manifestations and natural history of thoracic aortic aneurysms depend on their location. Cystic medial necrosis is the most common cause of ascending aortic aneurysms,

whereas atherosclerosis is the condition most frequently associated with aneurysms of the aortic arch and descending thoracic aorta. The average growth rate of thoracic aneurysms is 0.1 to 0.4 cm per year. The risk of rupture is related to the size of the aneurysm and the presence of symptoms; it increases substantially for ascending aortic aneurysms >6 cm and descending thoracic aneurysms >7 cm. Most thoracic aortic aneurysms are asymptomatic. However, compression or erosion of adjacent tissue by aneurysms may cause symptoms such as chest pain, shortness of breath, cough, hoarseness, or dysphagia. Aneurysmal dilatation of the ascending aorta may cause congestive heart failure as a consequence of aortic regurgitation; and compression of the superior vena cava may produce congestion of the head, neck, and upper extremities.

A chest x-ray may be the first test to suggest the diagnosis of a thoracic aortic aneurysm (Fig. 231-1). Findings include widening of the mediastinal shadow and displacement or compression of the trachea or left mainstem bronchus. Two-dimensional echocardiography, and particularly transesophageal echocardiography, can be used to assess the proximal ascending aorta and descending thoracic aorta. Both contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) are sensitive and specific tests for assessment of aneurysms of the thoracic aorta. In asymptomatic patients whose aneurysms are too small to justify surgery, noninvasive testing with either contrast-enhanced CT or MRI should be performed at least every 6 to 12 months to monitor expansion. Contrast aortography is frequently required preoperatively to assess the length of the aneurysm and involvement of branch vessels (Fig. 231-2).

FIGURE 231-1 A chest x-ray of a patient with a thoracic aortic aneurysm.

FIGURE 231-2 An aortogram demonstrating a large fusiform aneurysm of the descending thoracic aorta.

Patients with thoracic aortic aneurysms, and particularly patients with Marfan syndrome who have evidence of aortic root dilatation, should receive long-term beta-blocker therapy. Additional medical therapy should be given, as necessary, to control hypertension.

Operative repair with placement of a prosthetic graft is indicated in patients with symptomatic thoracic aortic aneurysms, and in those in whom the aortic diameter is >6 cm or has increased by >1 cm per year. In patients with Marfan syndrome, thoracic aortic aneurysms >5 cm should be considered for surgery.

ABDOMINAL AORTIC ANEURYSMS

Abdominal aortic aneurysms occur more frequently in males than in females, and the incidence increases with age. Abdominal aortic aneurysms may affect 1 to 2% of men older than 50 years. At least 90% of all abdominal aortic aneurysms >4.0 cm are affected by atherosclerosis, and most of these aneurysms are below the level of the renal arteries. Prognosis is related to both the size of the aneurysm and the severity of coexisting coronary artery and cerebrovascular disease. The risk of rupture increases with the size of the aneurysm. The 5-year risk of rupture for aneurysms <5 cm is 1 to 2%, whereas it is 20 to 40% for aneurysms >5 cm in diameter. The formation of mural thrombi within the aneurysm may predispose to peripheral embolization.

An abdominal aortic aneurysm commonly produces no symptoms. It is usually detected on routine examination as a palpable, pulsatile, and nontender mass, or it is an incidental finding during an abdominal x-ray or ultrasound performed for other reasons. However, as abdominal aortic aneurysms expand, they may become painful. Some patients complain of strong pulsations in the abdomen; others experience pain in the chest, lower back, or scrotum. Aneurysmal pain is usually a harbinger of rupture and represents a medical emergency. More often, acute rupture occurs without any prior warning, and this complication is always life-threatening. Rarely, there is leakage of the aneurysm with severe pain and tenderness. Acute pain and hypotension occur with rupture of the aneurysm, which requires emergency operation.

Abdominal radiography may demonstrate the calcified outline of the aneurysm. However, about 25% of aneurysms are not calcified and cannot be visualized by plain x-ray. An abdominal ultrasound can delineate the transverse and longitudinal dimensions of an abdominal aortic aneurysm and may detect mural thrombus. Abdominal ultrasound is useful for serial documentation of aneurysm size and can be used to screen patients at risk for developing aortic aneurysm, such as those with affected siblings, peripheral atherosclerosis, or peripheral artery aneurysms. In one larger study, ultrasound screening of men aged 65 to 74 years was associated with a risk reduction in aneurysm-related death by 42%. CT with contrast and MRI are accurate, noninvasive tests to determine the location and size of abdominal aortic aneurysms (Fig. 231-3). Contrast aortography is used for the evaluation of patients with aneurysms before surgery, but the procedure carries a small risk of complications, such as bleeding, allergic reactions, and atheroembolism. This technique is useful in documenting the length of the aneurysm, especially its upper and lower limits, and the extent of associated atherosclerotic vascular disease. However, since the presence of mural clots may reduce the luminal size, aortography may underestimate the diameter of an aneurysm.

FIGURE 231-3 A computed tomographic angiogram (CTA) depicting a fusiform abdominal aortic

aneurysm that has been treated with a bifurcated stent graft.

TREATMENT

Operative repair of the aneurysm and insertion of a prosthetic graft is indicated for abdominal aortic aneurysms of any size that are expanding rapidly or are associated with symptoms. For asymptomatic aneurysms, operation is indicated if the diameter is >5.5 cm. Operation may be recommended in patients with aneurysm diameters of 4 to 5 cm, except for patients with exceptionally high operative risk. However, in recent randomized trials of patients with abdominal aortic aneurysms <5.5 cm, there was no difference in the long-term (5- to 8-year) mortality rate between those followed with ultrasound surveillance and those undergoing elective aneurysm repair. Thus, serial noninvasive follow-up of smaller aneurysms (<5 cm) is an alternative to immediate surgery. Percutaneous placement of endovascular stent grafts (Fig. 231-3) for treatment of infrarenal abdominal aortic aneurysms is currently available for selected patients, and initial reports have been favorable.

In surgical candidates, careful preoperative cardiac and general medical evaluations (followed by appropriate therapy of complicating conditions) are essential. Preexisting coronary artery disease, congestive heart failure, pulmonary disease, diabetes, and advanced age add to the risk of surgery. Perioperative management should include the placement of a Swan-Ganz catheter and arterial line to monitor and optimize left ventricular filling pressure, cardiac output, and arterial pressure, especially during clamping and unclamping of the aorta, as well as during the immediate postoperative period. With careful preoperative cardiac evaluation and postoperative care, the operative mortality rate approximates 1 to 2%. After acute rupture, the mortality rate of emergent operation generally exceeds 50%.

AORTIC DISSECTION

Aortic dissection is caused by a circumferential or, less frequently, transverse tear of the intima. It often occurs along the right lateral wall of the ascending aorta where the hydraulic shear stress is high. Another common site is the descending thoracic aorta just below the ligamentum arteriosum. The initiating event is either a primary intimal tear with secondary dissection into the media or a medial hemorrhage that dissects into and disrupts the intima. The pulsatile aortic flow then dissects along the elastic lamellar plates of the aorta and creates a false lumen. The dissection usually propagates distally down the descending aorta and into its major branches, but it may also propagate proximally. In some cases, a secondary distal intimal disruption occurs, resulting in the reentry of blood from the false to the true lumen.

There are at least two important pathologic and radiologic variants: intramural hematoma

without an intimal flap and penetrating atherosclerotic ulcer. The clinical picture and therapeutic management of intramural hematoma are similar to those for classic aortic dissection. By contrast, penetrating ulcers are usually localized and are not associated with extensive propagation. They are primarily found in the mid and distal portions of the descending thoracic aorta and are associated with extensive atherosclerotic disease. The ulcer can erode beyond the intimal border, leading to medial hematoma, and may progress to false aneurysm formation or rupture.

DeBakey and coworkers initially classified aortic dissections as type I, in which an intimal tear occurs in the ascending aorta but which involves the descending aorta as well; type II, in which the dissection is limited to the ascending aorta; and type III, in which the intimal tear is located in the descending area with distal propagation of the dissection (Fig. 231-4). Another classification (Stanford) is that of type A, in which the dissection involves the ascending aorta (proximal dissection), and type B, in which it is limited to the descending aorta (distal dissection). From a management standpoint, classification into type A or B is more practical and useful, since DeBakey types I and II are managed in a similar manner.

FIGURE 231-4 Classification of aortic dissections. Stanford classification: Type A dissections (*top panels*) involve the ascending aorta independent of site of tear and distal extension; type B dissections (*bottom panels*) involve transverse and/or descending aorta without involvement of the ascending aorta. DeBakey classification: Type I dissection involves ascending to descending aorta (*top left*); type II dissection is limited to ascending or transverse aorta, without descending aorta (*top center + top right*); type III dissection involves descending aorta only (*bottom left*). *[From DC Miller, in RM Doroghazi, EE Slater (eds): Aortic Dissection. New York, McGraw-Hill, 1983, with permission.]*

The factors that predispose to aortic dissection include systemic hypertension, a coexisting condition in 70% of patients, and cystic medial necrosis. Aortic dissection is the major cause of morbidity and mortality in patients with Marfan syndrome (Chap. 342) and similarly may affect patients with Ehlers-Danlos syndrome. The incidence is

also increased in patients with inflammatory aortitis (i.e., Takayasu's arteritis, giant cell arteritis), congenital aortic valve anomalies (e.g., bicuspid valve), in those with coarctation of the aorta, and in otherwise normal women during the third trimester of pregnancy.

CLINICAL MANIFESTATIONS

The peak incidence is in the sixth and seventh decades. Men are more affected than women by a ratio of 2:1. The presentations of aortic dissection and its variants are the consequences of intimal tear, dissecting hematoma, occlusion of involved arteries, and compression of adjacent tissues. Acute aortic dissection presents with the sudden onset of pain (Chap. 12), which is often described as very severe and tearing and is associated with diaphoresis. The pain may be localized to the front or back of the chest, often the interscapular region, and typically migrates with propagation of the dissection. Other symptoms include syncope, dyspnea, and weakness. Physical findings may include hypertension or hypotension, loss of pulses, aortic regurgitation, pulmonary edema, and neurologic findings due to carotid artery obstruction (hemiplegia, hemianesthesia) or spinal

cord ischemia (paraplegia). Bowel ischemia, hematuria, and myocardial ischemia have all been observed. These clinical manifestations reflect complications resulting from the dissection occluding the major arteries. Furthermore, clinical manifestations may result from the compression of adjacent structures (e.g., superior cervical ganglia, superior vena cava, bronchus, esophagus) by the expanding dissection causing aneurysmal dilatation, and include Horner's syndrome, superior vena caval syndrome, hoarseness, dysphagia, and airway compromise. Hemopericardium and cardiac tamponade may complicate a type A lesion with retrograde dissection. Acute aortic regurgitation is an important and common (>50%) complication of proximal dissection. It is the outcome of either a circumferential tear that widens the aortic root or a disruption of the annulus by dissecting hematoma that tears a leaflet(s) or displaces it below the line of closure. Signs of aortic regurgitation include bounding pulses, a wide pulse pressure, a diastolic murmur often radiating along the right sternal border, and evidence of congestive heart failure. The clinical manifestation depends on the severity of the regurgitation.

In dissections involving the ascending aorta, the chest x-ray often reveals a widened superior mediastinum. A pleural effusion (usually left-sided) may also be present. This effusion is typically serosanguineous and not indicative of rupture unless accompanied by hypotension and falling hematocrit. In dissections of the descending thoracic aorta, a widened mediastinum may also be observed on chest x-ray. In addition, the descending aorta may appear to be wider than the ascending portion. An electrocardiogram that shows no evidence of myocardial ischemia is helpful in distinguishing aortic dissection from myocardial infarction. Rarely, the dissection involves the right or left coronary ostium and causes acute myocardial infarction. The diagnosis of aortic dissection can be established by aortography or by the use of noninvasive techniques such as echocardiography, CT, or MRI. Aortography may be used to document the diagnosis; to identify the entry point, the intimal flap, and the false and true lumina; and to establish the extent of dissection into the major arteries. Coronary angiography may be performed concomitantly in high-risk patients in the evaluation and preparation for surgery. The sensitivity of aortography is 70% for visualizing an intimal flap, 56% for the site of intimal tear, and 87% for false lumen. It is unable to recognize intramural hemorrhage. Transthoracic echocardiography can be performed simply and rapidly and has an overall sensitivity of 60 to 85%. For diagnosing proximal ascending aortic dissections, its sensitivity exceeds 80%; it is less useful for detecting dissection of the arch and descending thoracic aorta. Transesophageal echocardiography requires greater skill and patient cooperation but is very accurate in identifying dissections of the ascending and descending thoracic aorta, but not the arch, achieving 98% sensitivity and approximately 90% specificity. Echocardiography also provides important information regarding the presence and severity of aortic regurgitation and pericardial effusion. CT and MRI are both highly accurate in identifying the intimal flap and the extent of the dissection; each has a sensitivity and specificity >90%. They are useful in recognizing intramural hemorrhage and penetrating ulcers. MRI can also detect blood flow, which may be useful in characterizing antegrade versus retrograde dissection. Transesophageal echocardiography, CT, and MRI are the diagnostic procedures of choice over contrast aortography. Their relative utility depends on the availability and expertise in individual institutions as well as on the hemodynamic stability of the patient, with CT and MRI obviously less suitable for unstable patients.

TREATMENT

Medical therapy should be initiated as soon as the diagnosis is considered. The patient should be admitted to an intensive care unit for monitoring hemodynamics and urine output. Unless hypotension is present, therapy should be aimed at reducing cardiac contractility and systemic arterial pressure, and thereby shear stress. For acute dissection, unless contraindicated, β-adrenergic blockers should be administered parenterally, using intravenous propranolol, metoprolol, or the short-acting esmolol to achieve a heart rate of approximately 60 beats/min. This should be accompanied by sodium nitroprusside infusion to lower systolic blood pressure to ≤120 mmHg. Labetalol (Chap. 230), a drug with both β- and α-adrenergic blocking properties, is also used as a parenteral agent in the acute therapy of dissection.

The calcium channel antagonists, verapamil and diltiazem, may be used intravenously if nitroprusside or labetalol cannot be employed. The addition of a parenteral angiotensin-converting enzyme (ACE) inhibitor, such as enalaprilat, to a β-adrenergic blocker may also be considered. Isolated use of direct vasodilators, such as diazoxide and hydralazine, is contraindicated because these agents can increase hydraulic shear and may propagate dissection.

Emergent or urgent surgical correction is the preferred treatment for ascending aortic dissections (type A) and complicated type B dissections including those characterized by propagation, compromise of major aortic branches, impending rupture, or continued pain. Surgery involves excision of the intimal flap, obliteration of the false lumen, and placement of an interposition graft. A composite valve-graft conduit is used if the aortic valve is disrupted. The overall in-hospital mortality rate after surgical treatment of patients with aortic dissection is reported to be 15 to 25%. The major causes of perioperative mortality and morbidity include myocardial infarction, paraplegia, renal failure, tamponade, hemorrhage, and sepsis. Reports of the use of endoluminal stent grafts in selected patients with type B dissection are encouraging. Other transcatheter techniques, such as fenestration of the intimal flaps and stenting of narrowed branch vessels to increase flow to compromised organs, are also under investigation. For uncomplicated and stable distal dissection (type B), medical therapy is the preferred treatment. The in-hospital mortality rate of medically treated patients with type B dissection is 10 to 20%. Long-term therapy for patients with aortic dissection (with or without surgery) consists of the control of hypertension and reduction of cardiac contractility with the use of beta blockers plus other antihypertensive agents such as ACE inhibitors or calcium antagonists. Patients with chronic type B dissection should be followed on an outpatient basis every 6 to 12 months by contrast-enhanced CT or MRI to detect propagation or expansion. Patients with Marfan syndrome are at high risk for postdissection complications. The long-term prognosis for patients with treated dissections is generally good with careful follow-up; the 10-year survival rate is approximately 60%.

AORTIC OCCLUSION

CHRONIC ATHEROSCLEROTIC OCCLUSIVE DISEASE

Atherosclerosis may affect the thoracic and abdominal aorta. Occlusive aortic disease caused by atherosclerosis is usually confined to the distal abdominal aorta below the renal arteries. Frequently the disease extends to the iliac arteries (Chap. 232). Claudication characteristically involves the lower back, buttocks, and thighs and may be associated with impotence in males

(Leriche syndrome). The severity of the symptoms depends on the adequacy of collaterals. With sufficient collateral blood flow, a complete occlusion of the abdominal aorta may occur without the development of ischemic symptoms. The physical findings include absence of femoral and other distal pulses bilaterally and the detection of an audible bruit over the abdomen (usually at or below the umbilicus) and the common femoral arteries. Atrophic skin, loss of hair, and coolness of the lower extremities are usually observed. In advanced ischemia, rubor on dependency and pallor on elevation can be seen.

The diagnosis is usually established by the physical examination and noninvasive testing, including leg pressure measurements, Doppler velocity analysis, pulse volume recordings, and duplex ultrasonography. The anatomy may be defined by MRI, CT or conventional aortography before revascularization. Operative treatment is indicated in patients with debilitating symptoms and/or with the development of leg ischemia.

ACUTE OCCLUSION

Acute occlusion in the distal abdominal aorta represents a medical emergency because it threatens the viability of the lower extremities. It usually results from an occlusive embolus that almost always originates from the heart. Rarely, acute occlusion may occur as the result of in situ thrombosis in a preexisting severely narrowed segment of the aorta or plaque rupture and hemorrhage into such an area.

The clinical picture is one of acute ischemia of the lower extremities. Severe rest pain, coolness, and pallor of the lower extremities and the absence of distal pulses bilaterally are the usual manifestations. Diagnosis should be established rapidly by aortography. Emergency thrombectomy or revascularization is indicated.

AORTITIS

Aortitis frequently affects the thoracic aorta and may result in aneurysmal dilatation and aortic regurgitation; it occasionally obstructs branch vessels of the aorta.

SYPHILITIC AORTITIS

This late manifestation of luetic infection (Chap. 153) usually affects the proximal ascending aorta, particularly the aortic root, resulting in aortic dilatation and aneurysm formation. Syphilitic aortitis may occasionally involve the aortic arch or the descending aorta. The aneurysms may be saccular or fusiform and are usually asymptomatic, but compression of and erosion into adjacent structures may result in symptoms; rupture may

also occur.

The initial lesion is an obliterative endarteritis of the vasa vasorum, especially in the adventitia. This is an inflammatory response to the invasion of the adventitia by the spirochetes. Destruction of the aortic media occurs as the spirochetes spread into this layer, usually via the lymphatics accompanying the vasa vasorum. Destruction of collagen and elastic tissues leads to dilation of the aorta, scar formation, and calcification. These changes account for the characteristic radiographic appearance of a calcified ascending aortic aneurysm.

The disease typically presents as an incidental chest radiographic finding 15 to 30 years after initial infection. Symptoms may result from aortic regurgitation, narrowing of coronary ostia due to syphilitic aortitis, compression of adjacent structures (e.g., esophagus), or rupture. Diagnosis is established by a positive serologic test, i.e., rapid plasmin reagin (RPR) or fluorescent treponemal antibody. Treatment includes penicillin and surgical excision and repair.

RHEUMATIC AORTITIS

Rheumatoid arthritis (Chap. 301), ankylosing spondylitis (Chap. 305), psoriatic arthritis (Chap. 305), Reiter's syndrome (Chap. 305), relapsing polychondritis, and inflammatory bowel disorders may all be associated with aortitis involving the ascending aorta. The inflammatory lesions usually involve the ascending aorta and may extend to the sinuses of Valsalva, the mitral valve leaflets, and adjacent myocardium. The clinical manifestations are aneurysm, aortic regurgitation, and involvement of the cardiac conduction system.

TAKAYASU'S ARTERITIS

This inflammatory disease often affects the ascending aorta and aortic arch causing obstruction of the aorta and its major arteries. Takayasu's arteritis is also termed *pulseless disease* because of the frequent occlusion of the large arteries originating from the aorta. It may also involve the descending thoracic and abdominal aorta and occlude large branches such as the renal arteries. Aortic aneurysms may also occur. The pathology is a panarteritis, characterized by mononuclear cells and occasionally giant cells, with marked intimal hyperplasia, medial and adventitial thickening, and, in chronic form, fibrotic occlusion. The disease is most prevalent in young females of Asian descent but does occur in women of other geographic and ethnic origins and also in young men. During the acute stage, fever, malaise, weight loss, and other systemic symptoms may be evident. An elevation of the erythrocyte sedimentation rate is common. The chronic stages of the disease present with symptoms related to large artery occlusion, such as upper extremity claudication, cerebral ischemia, and syncope. The chronic disease is intermittently active. Since the process is progressive and there is no definitive therapy, the prognosis is usually poor. Glucocorticoids and immunosuppressive agents have been reported to be effective in some patients during the acute phase. Occasionally, anticoagulation prevents thrombosis and complete occlusion of a large artery. Surgical bypass or endovascular intervention of a critically stenotic artery may be necessary.

GIANT CELL ARTERITIS (See also Chap. 306*)*

This vasculitis occurs in older individuals and affects women more often than men. Primarily large and medium-sized arteries are affected. The pathology is that of focal granulomatous lesions involving the entire arterial wall. It may be associated with polymyalgia rheumatica. Obstruction of medium-sized arteries (e.g., temporal and ophthalmic arteries) and of major branches of the aorta and the development of aortitis and aortic regurgitation are some of the complications of the disease. High-dose glucocorticoid therapy may be effective when given early.

FURTHER READING

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