chapter 22

Thoracic Aneurysms and Aortic Dissection

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ANATOMY OF THE AORTA

The aorta consists of two major segments—the proximal aorta and the distal aorta—whose anatomic characteristics affect both the clinical manifestations of disease in these segments and the selection of treatment strategies for such disease (Fig. 22-1). The proximal aortic segment includes the ascending aorta and the transverse aortic arch. The ascending aorta begins at the aortic valve and ends at the origin of the innominate artery. The first portion of the ascending aorta is the aortic root, which includes the aortic valve annulus and the three sinuses of Valsalva; the coronary arteries originate from two of these sinuses. The aortic root joins the tubular portion of the ascending aorta at the sinotubular ridge. The transverse aortic arch is the area from which the brachiocephalic branches arise. The distal aortic segment includes the descending thoracic aorta and the abdominal aorta. The descending thoracic aorta begins distal to the origin of the left subclavian artery and extends to the diaphragmatic hiatus, where it joins the abdominal aorta. The descending thoracic aorta gives rise to multiple bronchial and esophageal branches, as well as to the segmental intercostal arteries, which provide circulation to the spinal cord.

The volume of blood that flows through the thoracic aorta at high pressure is far greater than that found in any other vascular structure. For this reason, any condition that disrupts the integrity of the thoracic aorta, such as aortic dissection, aneurysm rupture, or traumatic injury, can have catastrophic consequences.

Historically, open surgical repair of such conditions has been an intimidating undertaking associated with significant morbidity and mortality. Strategies for protecting the brain and spinal cord during such repairs have become critical in preventing devastating complications. Endovascular therapy for such conditions in selected patients has become accepted practice, producing fewer adverse outcomes than traditional approaches.

THORACIC AORTIC ANEURYSMS

Aortic aneurysm is defined as a permanent, localized dilatation of the aorta to a diameter that is at least 50% greater than is normal at that anatomic level.[1] The annual incidence of thoracic aortic aneurysms is estimated to be 5.9 per 100,000 persons.[2] The clinical manifestations, methods of treatment, and treatment results in patients with aortic aneurysms vary according to the cause and the aortic segment involved. Causes of thoracic aortic aneurysms include degenerative disease of the aortic wall, aortic dissection, aortitis, infection, and trauma. Aneurysms can be localized to a single aortic segment, or they can involve multiple segments. Thoracoabdominal aortic aneurysms, for example, involve both the descending thoracic aorta and the abdominal aorta. In the most extreme cases, the entire aorta is aneurysmal; this condition is often called *mega-aorta*.

Aortic aneurysms can be either "true" or "false." True aneurysms can take two forms: fusiform and saccular. Fusiform aneurysms are more common and can be described as symmetrical dilatations of the aorta. Saccular aneurysms are localized outpouchings of the aorta. False aneurysms, also called *pseudoaneurysms*, are leaks in the aortic wall that are contained by the outer layer of the aorta and/or the periaortic tissue; they are caused by disruption of the aortic wall and lead blood to collect in pouches of fibrotic tissue.

Aneurysms of the thoracic aorta consistently increase in size and eventually progress to cause serious complications. These include rupture, which usually is a fatal event. Therefore, aggressive treatment is indicated in all but the poorest surgical candidates. Small, asymptomatic thoracic aortic aneurysms can be followed, especially in high-surgical-risk patients, and can be treated surgically later if symptoms or complications develop, or if progressive enlargement occurs. Meticulous control of hypertension is the primary medical treatment for patients with small, asymptomatic aneurysms.

Elective resection with graft replacement is indicated in asymptomatic patients with an aortic diameter of at least twice

Key Points

1. Assessing urgency of repair is essential to developing the appropriate management plan. Although emergent repair carries greater operative risk than does elective repair, any inappropriate delay of repair risks death.
2. The clinical progression of an aortic aneurysm is continued expansion and eventual dissection or rupture. Hence, regular noninvasive imaging studies, as part of a lifelong surveillance plan, are necessary to ensure long-term patient health. Even small asymptomatic aneurysms should be routinely imaged to assess overall size and yearly rate of expansion.
3. Endovascular repair devices are approved for the treatment of descending thoracic aortic aneurysms, descending thoracic aortic dissections, aortic trauma, and penetrating aortic ulcer.
4. Practice guidelines have been published to help standardize the decision-making process and select an appropriate surgical intervention, as well as to standardize the use of imaging studies for patients with thoracic aortic disease.
5. Ascending aortic aneurysms that are symptomatic or ≥5.5 cm in diameter should be repaired regardless of whether the patient has a bicuspid or tricuspid aortic valve. This threshold is lowered for patients with certain heritable disorders affecting the aorta and for patients with additional risk factors, such as rapid aortic expansion (≥0.5 cm per year) or a family history of dissection.
6. Surgical repair involves the development of a patient-tailored plan based on careful preoperative medical evaluation. When appropriate, optimizing a patient's health status—to mitigate existing comorbidities—is important before surgical intervention.
7. The development and use of surgical adjuncts like antegrade selective cerebral perfusion and cerebrospinal fluid drainage have significantly reduced the morbidity rates traditionally associated with complex aortic repair.
8. Proximal aortic dissection is a life-threatening condition, and immediate operative repair is generally indicated, although definitive aortic repair may be delayed until after severe malperfusion has been treated.

normal in the involved segment (5 to 6 cm in most thoracic segments). Elective repair is contraindicated by extreme operative risk due to severe coexisting cardiac or pulmonary disease and by other conditions that limit life expectancy, such as malignancy. An emergency operation is performed for any patient in whom a ruptured aneurysm is suspected.

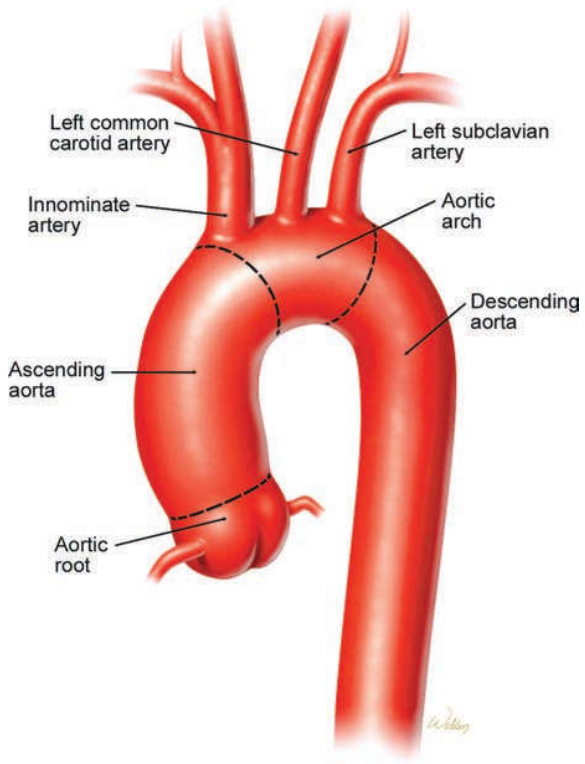


Figure 22-1. Illustration of normal thoracic aortic anatomy. The brachiocephalic vessels arise from the transverse aortic arch and are used as anatomic landmarks to define the aortic regions. The ascending aorta is proximal to the innominate artery, whereas the descending aorta is distal to the left subclavian artery.

Patients with thoracic aortic aneurysm often have coexisting aneurysms of other aortic segments. A common cause of death after repair of a thoracic aortic aneurysm is rupture of a different aortic aneurysm. Therefore, staged repair of multiple aortic segments often is necessary. As with any major operation, careful preoperative evaluation for coexisting disease and subsequent medical optimization are important for successful surgical treatment.

An alternative to traditional open repair of a descending thoracic aortic aneurysm is endovascular stent grafting. Certain anatomic criteria for use—such as a landing zone that includes at least 2 cm of landing zone of healthy aortic tissue proximal and distal to the targeted aneurysm—are preferable, but not absolutely necessary. Although few data on long-term outcomes have recently been published, endovascular repair of descending thoracic aortic aneurysm has become an accepted practice that produces excellent midterm results.

Causes and Pathogenesis

General Considerations. The normal aorta derives its elasticity and tensile strength from the medial layer, which contains approximately 45 to 55 lamellae of elastin, collagen, smooth muscle cells, and ground substance. Elastin content is highest within the ascending aorta, as would be expected because of its compliant nature, and decreases distally into the descending and abdominal aorta. Maintenance of the aortic matrix involves complex interactions among smooth muscle cells, macrophages, proteases, and protease inhibitors. Any alteration in this delicate balance can lead to aortic disease.

Thoracic aortic aneurysms have a variety of causes (Table 22-1). Although these disparate pathologic processes differ in biochemical and histologic terms, they share the final common pathway of progressive aortic expansion and eventual rupture.

Hemodynamic factors clearly contribute to the process of aortic dilatation. The vicious cycle of increasing diameter and increasing wall tension, as characterized by Laplace's law (tension = pressure × radius), is well established. Turbulent

Table 22-1

Causes of thoracic aortic aneurysm

Nonspecific medial degeneration
Aortic dissection
Heritable conditions
Marfan syndrome
Loeys-Dietz syndrome
Ehlers-Danlos syndrome
Turner syndrome
Familial thoracic aortic aneurysm
Aneurysms-osteoarthritis syndrome
Congenital bicuspid aortic valve
Bovine aortic arch
Poststenotic dilatation
Infection
Aortitis
Takayasu arteritis
Giant cell arteritis
Rheumatoid aortitis
Trauma (pseudoaneurysm)

blood flow also is recognized as a factor. Poststenotic aortic dilatation, for example, occurs in some patients with aortic valve stenosis or coarctation of the descending thoracic aorta. Hemodynamic derangements, however, are only one piece of a complex puzzle.

Atherosclerosis is commonly cited as a cause of thoracic aortic aneurysms. However, although atherosclerotic disease often is found in conjunction with aortic aneurysms, the notion that atherosclerosis is a distinct cause of aneurysm formation has been challenged. In most thoracic aortic aneurysms, atherosclerosis appears to be a coexisting process, rather than the underlying cause.

Research into the pathogenesis of abdominal aortic aneurysms has focused on the molecular mechanisms of aortic wall degeneration and dilatation.[3] For example, imbalances between proteolytic enzymes (e.g., matrix metalloproteinases) and their inhibitors contribute to abdominal aortic aneurysm formation. Building on these advances, current investigations are attempting to determine whether similar inflammatory and proteolytic mechanisms are involved in thoracic aortic disease, in hope of identifying potential molecular targets for pharmacologic therapy.

Nonspecific Medial Degeneration. Nonspecific medial degeneration is the most common cause of thoracic aortic disease. Histologic findings of mild medial degeneration, including fragmentation of elastic fibers and loss of smooth muscle cells, are expected in the aging aorta. However, an advanced, accelerated form of medial degeneration leads to progressive weakening of the aortic wall, aneurysm formation, and eventual dissection, rupture, or both. The underlying causes of medial degenerative disease remain poorly understood.

Aortic Dissection. An aortic dissection usually begins as a tear in the inner aortic wall, which initiates a progressive separation of the medial layers and creates two channels within the aorta. This event profoundly weakens the outer wall. As the most common catastrophe involving the aorta, dissection represents a major, distinct cause of thoracic aortic aneurysms and is discussed in detail in the second half of this chapter.

Heritable Conditions. Several heritable conditions cause thoracic aortic aneurysms. To better characterize these disorders, the National Institutes of Health (NIH) sponsored a longitudinal registry for individuals affected by genetically triggered thoracic aortic aneurysms and cardiovascular conditions (GenTAC) more than a decade ago.[4] The registry enrollment includes adults and children in 13 clinical categories, including Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, familial thoracic aortic aneurysms and dissections, aneurysms-osteoarthritis syndrome, and congenital bicuspid aortic valve.

Marfan Syndrome Marfan syndrome is an autosomal dominant genetic disorder characterized by a specific connective tissue defect that leads to aneurysm formation. The phenotype of patients with Marfan syndrome typically includes a tall stature, high palate, joint hypermobility, eye lens disorders, mitral valve prolapse, and aortic aneurysms. The aortic wall is weakened by fragmentation of elastic fibers and deposition of extensive amounts of mucopolysaccharides (a process previously called *cystic medial degeneration* or *cystic medial necrosis*). Patients with Marfan syndrome have a mutation in the fibrillin gene located on the long arm of chromosome 15. The traditional view has been that abnormal fibrillin in the extracellular matrix decreases connective tissue strength in the aortic wall and produces abnormal elasticity, which predisposes the aorta to dilatation from wall tension caused by left ventricular ejection impulses.[5] More recent evidence, however, shows that the abnormal fibrillin causes degeneration of the aortic wall matrix by increasing the activity of transforming growth factor beta (TGF-β).[6] Between 75% and 85% of patients with Marfan syndrome have dilatation of the ascending aorta and annuloaortic ectasia (dilatation of the aortic sinuses and annulus).[7] Marfan syndrome also is frequently associated with aortic dissection, and aortic complications are the most common cause of death among patients with Marfan syndrome.[8]

Loeys-Dietz Syndrome Loeys-Dietz syndrome is phenotypically distinct from Marfan syndrome. It is characterized as an aneurysmal syndrome with widespread systemic involvement. Loeys-Dietz syndrome is an aggressive, autosomal dominant condition that is distinguished by the triad of arterial tortuosity and aneurysms, hypertelorism (widely spaced eyes), and bifid uvula or cleft palate. It is caused by heterozygous mutations in the genes encoding TGF-β receptors.[9,10] Patients with Loeys-Dietz syndrome—including young children—are at increased risk of aortic rupture and aortic dissection; diameter-based thresholds of repair tend to be lower for patients with this syndrome than for patients with other heritable disorders.

Ehlers-Danlos Syndrome Ehlers-Danlos syndrome includes a spectrum of inherited disorders of collagen synthesis. The subtypes represent differing defective steps of collagen production. Vascular type Ehlers-Danlos syndrome is characterized by an autosomal dominant defect in type III collagen synthesis, which can have life-threatening cardiovascular manifestations. Spontaneous arterial rupture, usually involving the mesenteric vessels, is the most common cause of death in these patients. Thoracic aortic aneurysms and dissections are less commonly associated with Ehlers-Danlos syndrome, but when they do occur, they pose a particularly challenging surgical problem because of the reduced integrity of the aortic tissue.[11] An Ehlers-Danlos variant of periventricular heterotopia associated with joint and skin hyperextensibility and aortic dilation has been described as being caused by mutations in the gene encoding filamin A

(*FLNA*), an actin-binding protein that links the smooth muscle cell contractile unit to the cell surface.[12]

Familial Thoracic Aortic Aneurysm and Dissection Families without the heritable syndromes described earlier also can be affected by genetic conditions that cause thoracic aortic aneurysm. In fact, it is estimated that at least 20% of patients with thoracic aortic aneurysms and dissections have a genetic predisposition to them. The involved mutations are characterized by autosomal dominant inheritance with decreased penetrance and variable expression. The number of genes for which mutations have been identified as causes of familial thoracic aortic aneurysm and dissection is expanding rapidly; involved genes include those related to TGF-β receptors (*TGFBR1* and *TGFBR2*), TGF-β ligands (*TGFB2* and *TGFB3*), myosin (*MYH11* and *MYLK*), elastin (*ELN*), elastin microfibril interfacer 1 (*EMLIN1*), microfibril-associated glycoprotein 2 (*MFAP5*), fibrillin-2 (*FBN2*), fibulin-4 (*FBLN4*), lysyl oxidase (*LOX*), and α-smooth muscle cell actin (*ACTA2*).[3,13-16] *ACTA2* mutations are present in approximately 14% of families with familial thoracic aortic aneurysms and dissections.

Aneurysms-Osteoarthritis Syndrome Aneurysms-osteoarthritis syndrome is an autosomal dominant disorder characterized by aortic and arterial aneurysms, arterial tortuosity, aortic dissection, mild craniofacial abnormalities, and early-onset osteoarthritis. Aneurysms-osteoarthritis syndrome is caused by mutations in the gene encoding SMAD3, a transcription factor for TGF-β. Affected patients have a high incidence of aortic dissection, which often occurs in a mildly dilated aorta and causes sudden death.[17]

Congenital Bicuspid Aortic Valve Bicuspid aortic valve is the most common congenital malformation of the heart or great vessels, affecting up to 2% of Americans.[18] Compared to patients with a normal, trileaflet aortic valve, patients with bicuspid aortic valve have an increased incidence of ascending aortic aneurysm formation and, often, a more rapid rate of aortic enlargement.[19] The location of the fused leaflet, or raphe, may be predictive of aortic dilation and other abnormalities.[20] Fifty to 70% of adults with bicuspid aortic valve, but without significant valve dysfunction, have echocardiographically detectable aortic dilatation.[21,22] This dilatation usually is limited to the ascending aorta and root.[23] Dilation occasionally is found in the arch and only rarely in the descending or abdominal aorta. In addition, aortic dissection occurs 10 times more often in patients with bicuspid valves than in the general population.[24] Recent findings suggest that aneurysms associated with bicuspid aortic valve have a fundamentally different pathobiologic cause than aneurysms that occur in patients with trileaflet valves.[25]

Although the exact mechanism responsible for aneurysm formation in patients with bicuspid aortic valve remains unclear, evidence suggests that these patients have a congenital connective tissue abnormality that predisposes the aorta to medial degeneration.[25-31] For example, fibrillin-1 content is significantly lower and matrix metalloproteinase activity is significantly higher in the aortic media in patients with bicuspid aortic valve than in persons with a normal, tricuspid aortic valve.[25-27] Further, the process of medial degeneration in patients with bicuspid aortic valve may be exacerbated by the presence of chronic turbulent flow through the deformed valve.

Bovine Aortic Arch Bovine aortic arch—a common origin of the innominate and left common carotid arteries—has been considered a normal anatomic variant. Studies from Yale University have identified a higher prevalence of bovine aortic arch in patients with thoracic aortic disease; an association was found between this anomaly and a generalized increase in aortic aneurysmal disease (without any predisposition to a particular aortic region). However, bovine aortic arch was not associated distinctly with bicuspid aortic valve or aortic dissection, but with a higher mean aortic growth rate: 0.29 cm per year in patients with bovine aortic arch, compared with 0.09 cm per year in controls. Therefore, bovine aortic arch may be better characterized as a precursor of aortic aneurysm than as a simple normal anatomic variant.[32] Further studies are needed to delineate the underlying mechanism for this association.

Infection. Primary infection of the aortic wall resulting in aneurysm formation is rare. Although these lesions are termed *mycotic aneurysms*, the responsible pathogens usually are bacteria rather than fungi. Bacterial invasion of the aortic wall may result from bacterial endocarditis, endothelial trauma caused by an aortic jet lesion, or extension from an infected laminar clot within a preexisting aneurysm. The most common causative organisms are *Staphylococcus aureus, Staphylococcus epidermidis, Salmonella*, and *Streptococcus*.[33,34] Unlike most other causes of thoracic aortic aneurysms, which generally produce fusiform aneurysms, infection often produces saccular aneurysms located in areas of aortic tissue destroyed by the infectious process.

Although syphilis was once the most common cause of ascending aortic aneurysms, the advent of effective antibiotic therapy has made syphilitic aneurysms a rarity in developed nations. In other parts of the world, however, syphilitic aneurysms remain a major cause of morbidity and mortality. The spirochete *Treponema pallidum* causes an obliterative endarteritis of the vasa vasorum that results in medial ischemia and loss of the elastic and muscular elements of the aortic wall. The ascending aorta and arch are the most commonly involved areas. The emergence of HIV infection in the 1980s was associated with a substantial increase in the incidence of syphilis in both HIV-positive and HIV-negative patients. Because syphilitic aortitis often presents 10 to 30 years after the primary infection, the incidence of associated aneurysms may increase in the near future.

Aortitis. In patients with preexisting degenerative thoracic aortic aneurysms, localized transmural inflammation and subsequent fibrosis can develop. The dense aortic infiltrate responsible for the fibrosis consists of lymphocytes, plasma cells, and giant cells. The cause of the intense inflammatory reaction is unknown. Although the severe inflammation is a superimposed problem rather than a primary cause, its onset within an aneurysm can further weaken the aortic wall and precipitate expansion.

Systemic autoimmune disorders also cause thoracic aortitis. Aortic Takayasu arteritis generally produces obstructive lesions related to severe intimal thickening, but associated medial necrosis can lead to aneurysm formation. In patients with giant cell arteritis (temporal arteritis), granulomatous inflammation may develop that involves the entire thickness of the aortic wall, causing intimal thickening and medial destruction. Rheumatoid aortitis is an uncommon systemic disease that is associated with rheumatoid arthritis and ankylosing spondylitis. The resulting medial inflammation and fibrosis can affect the aortic root, causing annular dilatation, aortic valve regurgitation, and ascending aortic aneurysm formation.

Pseudoaneurysms. Pseudoaneurysms of the thoracic aorta usually represent chronic leaks that are contained by surrounding

tissue and fibrosis. By definition, the wall of a pseudoaneurysm is not formed by intact aortic tissue; rather, the wall develops from organized thrombus and associated fibrosis. Pseudoaneurysms can arise from primary defects in the aortic wall (e.g., after trauma or contained aneurysm rupture) or from anastomotic or cannulation site leaks that occur after cardiovascular surgery. Anastomotic pseudoaneurysms can be caused by technical problems or by deterioration of the native aortic tissue, graft material, or suture. Commonly, they occur in patients with Marfan syndrome, Loeys-Dietz syndrome, or other heritable conditions that markedly weaken the vessel wall.[35] Tissue deterioration usually is related to either progressive degenerative disease or infection. Improvements in sutures, graft materials, and surgical techniques have decreased the incidence of thoracic aortic pseudoaneurysm. Should thoracic aortic pseudoaneurysms occur, they typically require expeditious open surgical or catheter-based repair because they are associated with a high incidence of morbidity and rupture.

Clinical History

Treatment decisions in cases of thoracic aortic aneurysm are guided by our current understanding of the clinical history of these aneurysms, which classically is characterized as progressive aortic dilatation and eventual dissection, rupture, or both. 1▶ An analysis by Elefteriades of data from 1600 patients with thoracic aortic disease has helped quantify these well-recognized risks.[36] Average expansion rates were 0.07 cm per year in ascending aortic aneurysms and 0.19 cm per year in descending thoracic aortic aneurysms. As expected, aortic diameter was a strong predictor of rupture, dissection, and mortality. For thoracic aortic aneurysms >6 cm in diameter, annual rates of catastrophic complications were 3.6% for rupture, 3.7% for dissection, and 10.8% for death. Critical "hinge-point" diameters, at which the incidence of expected complications significantly increased, were 6.0 cm for aneurysms of the ascending aorta and 7.0 cm for aneurysms of the descending thoracic aorta; the corresponding risks of rupture after reaching these diameters were 31% and 43%, respectively.[37]

Certain types of aneurysms have an elevated propensity for expansion and rupture. For example, aneurysms in patients with Marfan or Loeys-Dietz syndrome tend to dilate at an accelerated rate and rupture or dissect at smaller diameters than sporadic, nonheritable aneurysms. Before the era of surgical treatment for aortic aneurysms, the aggressive form of aortic disease in Marfan patients resulted in an average life expectancy of 32 years, with aortic root complications causing the majority of deaths.[38] Saccular aneurysms, which commonly are associated with aortic infection and typically affect only a discrete small section of the aorta, tend to grow more rapidly than fusiform aneurysms, which are associated with more widespread degenerative changes and generally affect a larger section of the aorta.

One common clinical scenario deserves special attention. A moderately dilated ascending aorta (i.e., 4 to 5 cm) often is encountered during aortic valve replacement or coronary artery bypass operations. The clinical history of these ectatic ascending aortas has been defined by several studies. Michel and colleagues[39] studied patients whose ascending aortic diameters were >4 cm at the time of aortic valve replacement; 25% of these patients required reoperation for ascending aortic replacement. Prenger and colleagues[40] reported that aortic dissection occurred in 27% of patients who had aortic diameters of >5 cm at the time of aortic valve replacement. Attention has been directed toward whether or not a mildly dilated aortic root should be replaced in patients with bicuspid aortic valve who are undergoing isolated valve replacement, and at what threshold to intervene. Although this is a controversial issue, many surgeons believe that the tendency toward late aortic dilatation in these patients warrants aggressive treatment.[41,42] According to a recent guidelines clarification,[43] in patients with bicuspid aortic valve who are undergoing aortic valve replacement or repair, replacing the ascending aorta is reasonable when the diameter of the ascending aorta is greater than 4.5 cm (Class IIa, Level C recommendation).

Clinical Manifestations

In many patients with thoracic aortic aneurysms, the aneurysm is discovered incidentally when imaging studies are performed for unrelated reasons. Therefore, patients often are asymptomatic at the time of diagnosis. However, thoracic aortic aneurysms that initially go undetected eventually create symptoms and signs that correspond with the segment of aorta that is involved. These aneurysms have a wide variety of manifestations, including compression or erosion of adjacent structures, aortic valve regurgitation, distal embolism, and rupture.

Local Compression and Erosion. Initially, aneurysmal expansion and impingement on adjacent structures causes mild, chronic pain. The most common symptom in patients with ascending aortic aneurysms is anterior chest discomfort; the pain is frequently precordial in location but may radiate to the neck and jaw, mimicking angina. Aneurysms of the ascending aorta and transverse aortic arch can cause symptoms related to compression of the superior vena cava, the pulmonary artery, the airway, or the sternum. Rarely, these aneurysms erode into the superior vena cava or right atrium, causing acute high-output failure. Expansion of the distal aortic arch can stretch the recurrent laryngeal nerve, which results in left vocal cord paralysis and hoarseness. Descending thoracic and thoracoabdominal aneurysms frequently cause back pain localized between the scapulae. When the aneurysm is largest in the region of the aortic hiatus, it may cause middle back and epigastric pain. Thoracic or lumbar vertebral body erosion typically causes severe, chronic back pain; extreme cases can present with spinal instability and neurologic deficits from spinal cord compression. Although mycotic aneurysms have a peculiar propensity to destroy vertebral bodies, spinal erosion also occurs with degenerative aneurysms. Descending thoracic aortic aneurysms may cause varying degrees of airway obstruction, manifesting as cough, wheezing, stridor, or pneumonitis. Pulmonary or airway erosion presents as hemoptysis. Compression and erosion of the esophagus cause dysphagia and hematemesis, respectively. Thoracoabdominal aortic aneurysms can cause duodenal obstruction or, if they erode through the bowel wall, gastrointestinal bleeding. Jaundice due to compression of the liver or porta hepatis is uncommon. Erosion into the inferior vena cava or iliac vein presents with an abdominal bruit, widened pulse pressure, edema, and heart failure.

Aortic Valve Regurgitation. Ascending aortic aneurysms can cause displacement of the aortic valve commissures and annular dilatation. The resulting deformation of the aortic valve leads to progressively worsening aortic valve regurgitation. In response to the volume overload, the heart remodels and becomes increasingly dilated. Patients with this condition may present with progressive heart failure, a widened pulse pressure, and a diastolic murmur.

Distal Embolization. Thoracic aortic aneurysms—particularly those involving the descending and thoracoabdominal aorta—are commonly lined with friable, atheromatous plaque and

mural thrombus. This debris may embolize distally, causing occlusion and thrombosis of the visceral, renal, or lower-extremity branches.

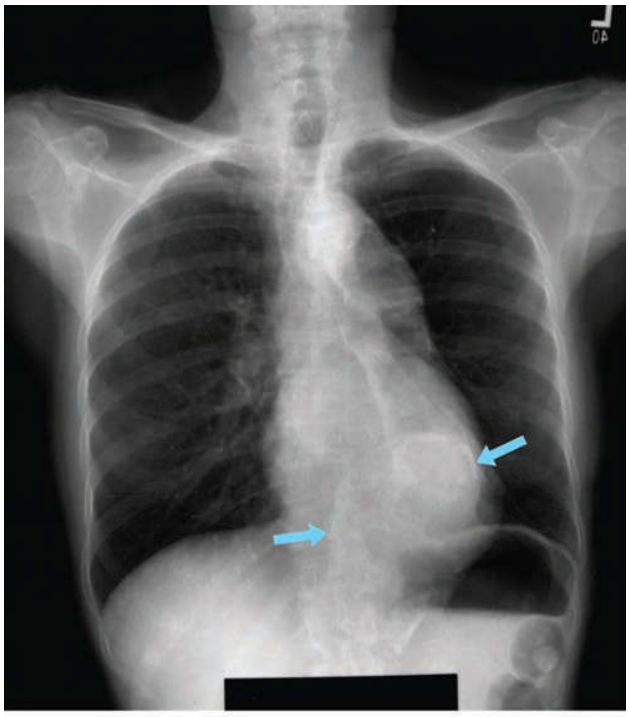
Rupture. Patients with ruptured thoracic aortic aneurysms often experience sudden, severe pain in the anterior chest (ascending aorta), upper back or left chest (descending thoracic aorta), or left flank or abdomen (thoracoabdominal aorta). When ascending aortic aneurysms rupture, they usually bleed into the pericardial space, producing acute cardiac tamponade and death. Descending thoracic aortic aneurysms rupture into the pleural cavity, producing a combination of severe hemorrhagic shock and respiratory compromise. External rupture is extremely rare; saccular syphilitic aneurysms have been observed to rupture externally after eroding through the sternum.

Diagnostic Evaluation

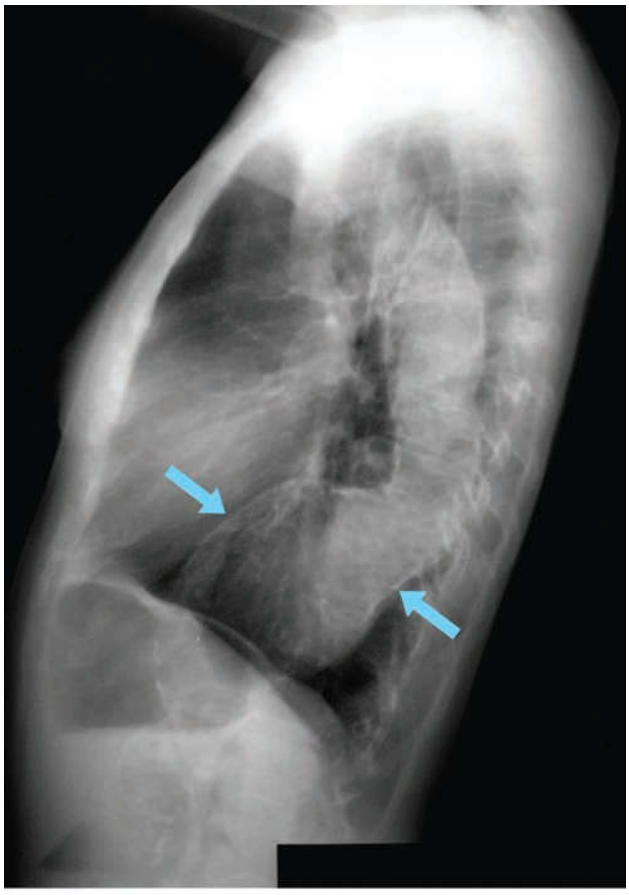
Diagnosis and characterization of thoracic aneurysms require imaging studies, which also provide critical information that guides the selection of treatment options. Although the best choice of imaging technique for the thoracic and thoracoabdominal aorta is somewhat institution-specific, varying with the availability of imaging equipment and expertise, efforts have been made to standardize key elements of image acquisition and reporting. Recent practice guidelines[44] recommend that aortic imaging reports plainly state the location of aortic abnormalities (including calcification and the extent to which abnormalities extend into branch vessels), the maximum external aortic diameters (rather than internal, lumen-based diameters), internal filling defects, and any evidence of rupture. Whenever possible, all results should be compared with those of prior imaging studies.

Plain Radiography. Plain radiographs of the chest, abdomen, or spine often provide enough information to support the initial diagnosis of thoracic aortic aneurysm. Ascending aortic aneurysms produce a convex shadow to the right of the cardiac silhouette. The anterior projection of an ascending aneurysm results in the loss of the retrosternal space in the lateral view. An aneurysm may be indistinguishable from elongation and tortuosity.[45] Importantly, chest radiographs (CXRs) may appear normal in patients with thoracic aortic disease and thus cannot exclude the diagnosis of aortic aneurysm. Aortic root aneurysms, for example, often are hidden within the cardiac silhouette. Plain CXRs may reveal convexity in the right superior mediastinum, loss of the retrosternal space, or widening of the descending thoracic aortic shadow, which may be highlighted by a rim of calcification outlining the dilated aneurysmal aortic wall. Aortic calcification also may be seen in the upper abdomen on a standard radiograph made in the anteroposterior or lateral projection (Fig. 22-2). Once a thoracic aortic aneurysm is detected on plain radiographs, additional studies are required to define the extent of aortic involvement.

Echocardiography and Abdominal Ultrasonography. Ascending aortic aneurysms are commonly discovered during echocardiography in patients presenting with symptoms or signs of aortic valve regurgitation. Both transthoracic and transesophageal echocardiography provide excellent visualization of the ascending aorta, including the aortic root.[46] Transesophageal echocardiography also allows visualization of the descending thoracic aorta but is not ideal for evaluating the transverse aortic arch (which is obscured by air in the tracheobronchial tree) or the upper abdominal aorta. Effective echocardiography requires considerable technical skill, both in obtaining adequate images and in interpreting them. This imaging modality has the added



A



B

Figure 22-2. Chest radiographs showing a calcified rim (*arrows*) in the aortic wall of a thoracoabdominal aortic aneurysm. **A.** Anteroposterior view. **B.** Lateral view.

benefit of assessing cardiac function and revealing any other abnormalities that may be present. During ultrasound evaluation of a suspected infrarenal abdominal aortic aneurysm, if a definitive neck cannot be identified at the level of the renal arteries, the possibility of thoracoabdominal aortic involvement should be suspected and investigated by using other imaging modalities. Caution should be exercised while interpreting aneurysm dimensions from ultrasound imaging because intraluminal measurements are often reported, whereas external measurements are usually used in other imaging modalities.

Computed Tomography. Computed tomographic (CT) scanning is widely available, provides visualization of the entire thoracic and abdominal aorta, and permits multiplanar and 3-dimensional aortic reconstructions. Consequently, CT is the most common—and arguably the most useful—imaging modality for evaluating thoracic aortic aneurysms.[47] In addition to establishing the diagnosis, CT provides information about an aneurysm's location, extent, anatomic anomalies, and relationship to major branch vessels. CT is particularly useful in determining the absolute diameter of the aorta, especially in the presence of laminated clot, and also detects aortic calcification. Contrast-enhanced CT provides information about the aortic lumen and can detect mural thrombus, aortic dissection, inflammatory periaortic fibrosis, and mediastinal or retroperitoneal hematoma due to contained aortic rupture. To increase consistency and ensure uniform reporting, current practice guidelines suggest that measurements be taken perpendicular to blood flow and at standard anatomic locations[44] (Fig. 22-3); this should reduce the likelihood of erroneous measurements, especially during serial imaging surveillance .

2▶

The major disadvantage of contrast-enhanced CT scanning is the possibility of contrast-induced acute renal failure in patients who are at risk (e.g., patients with preexisting renal disease or diabetes) even though the risk is smaller than was assumed in the past.[48,49] If possible, surgery is performed at least 1 day after contrast administration to allow time to observe renal function and to permit diuresis. If renal insufficiency occurs or is worsened, elective surgery is postponed until renal function returns to normal or stabilizes.

Magnetic Resonance Angiography. Magnetic resonance angiography (MRA) is becoming widely available and can facilitate visualization of the entire aorta. This modality produces aortic images comparable to those produced by contrast-enhanced CT but does not necessitate exposure to ionizing radiation.[50] In addition, MRA offers excellent visualization of branch-vessel details, and it is useful in detecting branch-vessel stenosis.[51] However, MRA is limited by high expense and a susceptibility to artifacts created by ferromagnetic materials, and gadolinium—the contrast agent for MRA—may be linked to nephrogenic systemic fibrosis and acute renal failure in patients with advanced renal insufficiency.[52] Furthermore, the MRA environment is not appropriate for many critically ill patients, and unlike CT imaging, MRA imaging is suboptimal in patients with extensive aortic calcification.

Invasive Aortography and Cardiac Catheterization. Although catheter-based contrast aortography was previously considered the gold standard for evaluating thoracic aortic disease, cross-sectional imaging (i.e., CT and MRA) has largely replaced this modality. Technologic improvements have enabled CT and MRA to provide excellent aortic imaging while causing less morbidity than catheter-based studies do, so CT and

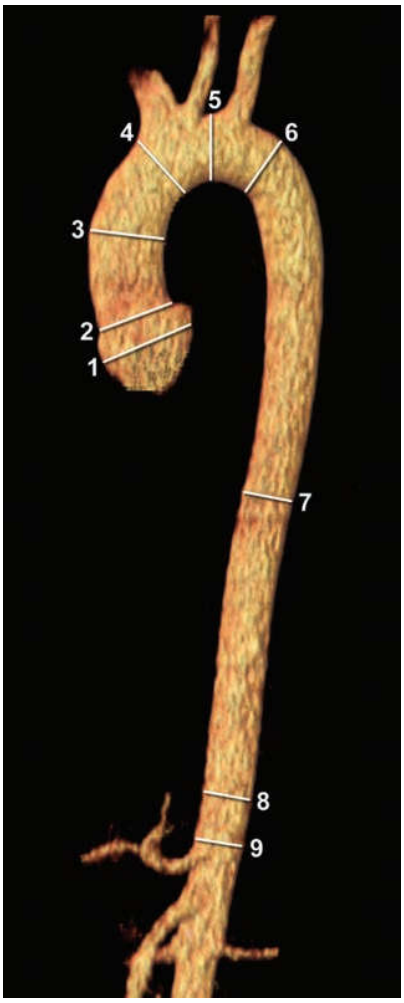


Figure 22-3. Current practice guidelines[44] seek to standardize the reporting of aortic diameters by indicating key locations of measurement. These include (1) the sinuses of Valsalva, (2) the sinotubular junction, (3) the mid-ascending aorta, (4) the proximal aortic arch at the origins of the innominate artery, (5) the mid-aortic arch, which is between the left common carotid and left subclavian arteries, (6) the proximal descending thoracic aorta, which begins at the isthmus (approximately 2 cm distal to the origins of the left subclavian artery), (7) the mid-descending thoracic artery, (8) the aorta at the diaphragm, and (9) the abdominal aorta at the origins of the celiac axis. *(Used with permission of Baylor College of Medicine.)*

MRA are now the primary modes for evaluating thoracic aortic disease. Today, the use of invasive aortography in patients with thoracic aortic disease is generally limited to those undergoing endovascular therapies or when other types of studies are contraindicated or have not provided satisfactory results.

Unlike standard aortography, cardiac catheterization continues to play an important role in diagnosis and preoperative planning, especially in patients with ascending aortic involvement. Proximal aortography can reveal not only the status of the coronary arteries and left ventricular function but also the degree of aortic valve regurgitation, the extent of aortic root involvement, coronary ostial displacement, and the relationship of the aneurysm to the arch vessels.

The value of the information one can obtain from catheter-based diagnostic studies should be weighed against

the established limitations and potential complications of such studies. A key limitation of aortography is that it images only the lumen and may therefore underrepresent the size of large aneurysms that contain laminated thrombus. Manipulation of intraluminal catheters can result in embolization of laminated thrombus or atheromatous debris. Proximal aortography carries a 0.6% to 1.2% risk of stroke. Other risks include allergic reaction to the contrast agent, iatrogenic aortic dissection, and bleeding at the arterial access site. In addition, the volumes of contrast agent required to adequately fill large aneurysms can cause significant renal toxicity. To minimize the risk of contrast nephropathy, patients receive periprocedural intravenous (IV) fluids for hydration, mannitol for diuresis, and acetylcysteine.[53,54] As with contrast-enhanced CT, surgery is performed ≥1 day after angiography whenever possible to ensure that renal function has stabilized or returned to baseline.

Treatment

Selecting the Appropriate Treatment. Once a thoracic aortic aneurysm is detected, management begins with patient education, particularly if the patient is asymptomatic, because aortic disease may progress rapidly and unexpectedly in some patients. A detailed medical history is collected, a physical examination is performed, and a systematic review of medical records is carried out to clearly assess the presence or absence of pertinent symptoms and signs, despite any initial denial of symptoms by the patient. Signs of heritable conditions such as Marfan syndrome or Loeys-Dietz syndrome are thoroughly reviewed. If clinical criteria are met for a heritable condition, confirmatory laboratory tests are conducted. Patients with heritable disorders are best treated in a dedicated aortic clinic where they can be appropriately followed up. Surveillance imaging and aggressive blood pressure control are the mainstays of initial management for asymptomatic patients. When patients become symptomatic or their aneurysms grow to meet certain size criteria, the patients become surgical candidates.

Endovascular therapy has become an accepted treatment for descending thoracic aortic aneurysm.[55,56] Its role in treating proximal aortic disease and thoracoabdominal aortic aneurysm remains experimental;[55] nonetheless, endoluminal stenting is approved by the U.S. Food and Drug Administration for the treatment of isolated descending thoracic aortic aneurysm, and several different devices have been approved for the treatment of blunt aortic injury and penetrating aortic ulcer. In practice, 3▶ however, the off-label application of aortic stent grafts is widespread and accounts for well over half their use[57]; endovascular approaches may be helpful in emergent aneurysm repair, such as for patients with aortic rupture.[58] Endovascular therapy has evolved to include hybrid repairs, which combine open "debranching" techniques (to reroute branching vessels) with endovascular aortic repair.[59,60] Despite these advances, for the repair of aneurysms with proximal aortic involvement and of thoracoabdominal aortic aneurysms, open procedures remain the gold standard and preferred approach.

Determination of the Extent and Severity of Disease. Cross-sectional imaging with reconstruction is critical when one is evaluating a thoracic aneurysm, determining treatment strategy, and planning necessary procedures. Note that patients with a thoracic aortic aneurysm may also have a second, remote aneurysm.[2] In such cases, the more threatening lesion usually is addressed first. In many patients, staged operative procedures are necessary for complete repair of extensive aneurysms involving the ascending aorta, transverse arch, and descending thoracic or thoracoabdominal aorta.[61] When the descending segment is not disproportionately large (compared with the proximal aorta) and is not causing symptoms, the proximal aortic repair is carried out first. An important benefit of this approach is that it allows treatment of valvular and coronary artery occlusive disease at the first operation.

Proximal aneurysms (proximal to the left subclavian artery) usually are addressed via a sternotomy approach. Aneurysms involving the descending thoracic aorta are evaluated in terms of criteria (described in the following section) for potential endovascular repair; those unsuitable for an endovascular approach are repaired with open techniques through a left thoracotomy. A CT scan can reveal detailed information about aortic calcification and luminal thrombus. These details are important in preventing embolization during surgical manipulation.

Indications for Operation Thoracic aortic aneurysms are repaired to prevent fatal rupture. Therefore, on the basis of clinical history studies and other data, practice guidelines for thoracic aortic disease[43,44,62] recommend elective operation in 4▶ asymptomatic patients when the diameter of an ascending aortic aneurysm is >5.5 cm, when the diameter of a descending thoracic aortic aneurysm is >6.0 cm, or when the rate of dilatation is >0.5 cm per year. In patients with heritable disorders such as Marfan and Loeys-Dietz syndromes, the threshold for operation is based on a smaller aortic diameter (5.0 cm for the ascending aorta in patients with Marfan syndrome, 4.4 to 4.6 cm for the ascending aorta in patients with Loeys-Dietz syndrome, and <6.0 cm for the descending thoracic aorta in patients with either disorder). For women with heritable disorders who are considering pregnancy, prophylactic aortic root replacement is 5▶ considered because the risk of aortic dissection or rupture increases at an aortic diameter of 4.0 cm and greater. For patients with ascending aortic aneurysm and bicuspid aortic valve, repair is recommended if aortic diameter is 5.0 cm or greater and additional risk factors are present (e.g., family history of dissection, expansion rate exceeding 0.5 cm per year), if aortic diameter is 5.5 cm or larger and no additional risk factors are present, or if aortic diameter exceeds 4.5 cm and the patient is undergoing aortic valve replacement or repair.[43] For low-risk patients with chronic aortic dissection, descending thoracic repair is recommended at an aortic diameter of 5.5 cm or greater.

The acuity of presentation is a major factor in decisions about the timing of surgical intervention. Many patients are asymptomatic at the time of presentation, so there is time for thorough preoperative evaluation and improvement of their current health status, such as through smoking cessation and other optimization programs. In contrast, patients who present with 6▶ symptoms may need urgent operation. Symptomatic patients are at increased risk of rupture and warrant expeditious evaluation. The onset of new pain in patients with known aneurysms is especially concerning because it may herald significant expansion, leakage, or impending rupture. Emergent intervention is reserved for patients who present with aneurysm rupture or superimposed acute dissection.[63]

Open Repair vs. Endovascular Repair As noted earlier, endovascular repair has become the standard approach for patients with isolated degenerative descending thoracic aortic aneurysm; in fact, practice guidelines recommend that endovascular repair be strongly considered for patients with descending thoracic aneurysm at an aortic diameter of 5.5 cm (which is slightly below the 6.0-cm threshold for open repair).[44] For endovascular

repairs to produce optimal outcomes, several anatomic criteria must be met. For one, the proximal and distal neck diameters should fall within a range that will allow proper sealing. Also, the proximal and distal landing zones should ideally be at least 20 mm long so that an appropriate seal can be made. Note that the limiting structures proximally and distally are the brachiocephalic vessels and celiac axis, respectively. Vascular access continues to be one of the most important determinants of successful deployment of the current endovascular devices. The femoral and iliac arteries have to be wide enough to accommodate the sheaths used to deploy the stent grafts. As endovascular technology evolves, newer devices are using smaller sheaths (or are "sheathless" self-deployed stent grafts) to accommodate smaller arteries. Tortuosity of the iliac vessels and abdominal aorta can make these procedures technically challenging. Occasionally, an 8- or 10-mm polyester "side graft" is anastomosed to the iliac artery through a retroperitoneal incision if the femoral vessels are too small to access easily.

Of note, attempts have been made to extend the use of endovascular therapy to aortic arch aneurysms and thoracoabdominal aortic aneurysms. Although reports of purely endovascular repair of the aortic arch remain limited, Greenberg and colleagues[64] have reported their experience with a large series of purely endovascular thoracoabdominal aortic repairs. Additionally, there have been numerous reports of small series of off-label, experimental hybrid procedures that involve debranching the aortic arch or the visceral vessels of the abdominal aorta, followed by endovascular exclusion of the aneurysm. The majority of hybrid approaches involve repairing the aortic arch.[59,60] In its simplest form, hybrid arch repair involves an open bypass from the left subclavian to the left common carotid artery, which is followed by deliberate coverage of the origins of the left subclavian artery by the stent graft. In its most complex form, hybrid arch repair involves rerouting all of the brachiocephalic vessels, followed by proximal placement of the stent graft in the ascending aorta and extending repair distally into the aortic arch and descending thoracic aorta.

The patients who theoretically benefit the most from an endovascular approach are those who are of advanced age or have significant comorbidities, as many of these patients face substantial risks when undergoing traditional open repair.[65] For example, with regard to open repair of a descending thoracic aortic aneurysm, significant pulmonary morbidity can occur postoperatively; therefore, patients with borderline pulmonary reserve may better tolerate an endovascular procedure than a standard open repair. Patients with heritable syndromic conditions generally are not considered candidates for elective endovascular repair except in specific circumstances.[66] Endovascular repair in patients with heritable syndromic conditions have produced poor results, which are mainly due to progressive dilatation, stent graft migration, and endoleak.[67,68]

Preoperative Assessment and Preparation. Given the impact of comorbid conditions on perioperative complications, a careful preoperative assessment of physiologic reserve is critical in assessing operative risk. Therefore, most patients undergo a thorough evaluation—with emphasis on cardiac, pulmonary, and renal function—before undergoing elective surgery.[69,70]

Cardiac Evaluation Coronary artery disease is common in patients with thoracic aortic aneurysm and is responsible for a substantial proportion of early and late postoperative deaths in such patients. Similarly, valvular disease and myocardial dysfunction have important implications when one is planning anesthetic management and surgical approaches for aortic repair. Transthoracic echocardiography is a satisfactory noninvasive method for evaluating both valvular and biventricular function. Dipyridamole-thallium myocardial scanning identifies regions of myocardium that have reversible ischemia, and this test is more practical than exercise testing in older patients with concomitant lower-extremity peripheral vascular disease. Cardiac catheterization and coronary arteriography are performed in patients who have evidence of coronary disease—as indicated by either the patient's history or the results of noninvasive studies—or who have a left ventricular ejection fraction of ≤30%. If significant valvular or coronary artery disease is identified before a proximal aortic operation, the disease can be addressed directly during the procedure. Patients who have asymptomatic distal aortic aneurysms and severe coronary occlusive disease undergo percutaneous transluminal angioplasty or surgical revascularization before the aneurysmal aortic segment is replaced.

Pulmonary Evaluation Pulmonary function screening with arterial blood gas measurement and spirometry is routinely performed before thoracic aortic operations. Patients with a forced expiratory volume in 1 second of >1.0 L and a partial pressure of carbon dioxide of <45 mmHg are considered appropriate candidates for open surgical repair. In suitable patients, borderline pulmonary function can be improved by implementing a regimen that includes smoking cessation, weight loss, exercise, and treatment of bronchitis for a period of 1 to 3 months before surgery. Although surgery is not withheld from patients with symptomatic aortic aneurysms and poor pulmonary function, adjustments in operative technique should be made to maximize these patients' chances of recovery. In such patients, preserving the left recurrent laryngeal nerve, the phrenic nerves, and diaphragmatic function is particularly important.

Renal Evaluation Renal function is assessed preoperatively by measuring serum electrolyte, blood urea nitrogen, and creatinine levels. Information about kidney size and perfusion can be obtained from the imaging studies used to evaluate the aorta.

Obtaining accurate information about baseline renal function has important therapeutic and prognostic implications. For example, perfusion strategies and perioperative medications are adjusted according to renal function. Patients with severely impaired renal function frequently require at least temporary hemodialysis after surgery. These patients also have a mortality rate that is significantly higher than normal. Patients with thoracoabdominal aortic aneurysms and poor renal function secondary to severe proximal renal occlusive disease undergo renal artery endarterectomy, stenting, or bypass grafting during the aortic repair.

Operative Repair

Proximal Thoracic Aortic Aneurysms

Open Repair Traditional open operations to repair proximal aortic aneurysms—which involve the ascending aorta, transverse aortic arch, or both—are performed through a midsternal incision and require cardiopulmonary bypass. The best choice of aortic replacement technique depends on the extent of the aneurysm and the condition of the aortic valve.[71] The spectrum of operations (Fig. 22-4) ranges from simple graft replacement of the tubular portion of the ascending aorta only (Fig. 22-4A) to replacement of the ascending aorta and the proximal aortic arch (Fig. 22-4B) to graft replacement of the entire proximal aorta, including the aortic root, and reattachment of the coronary

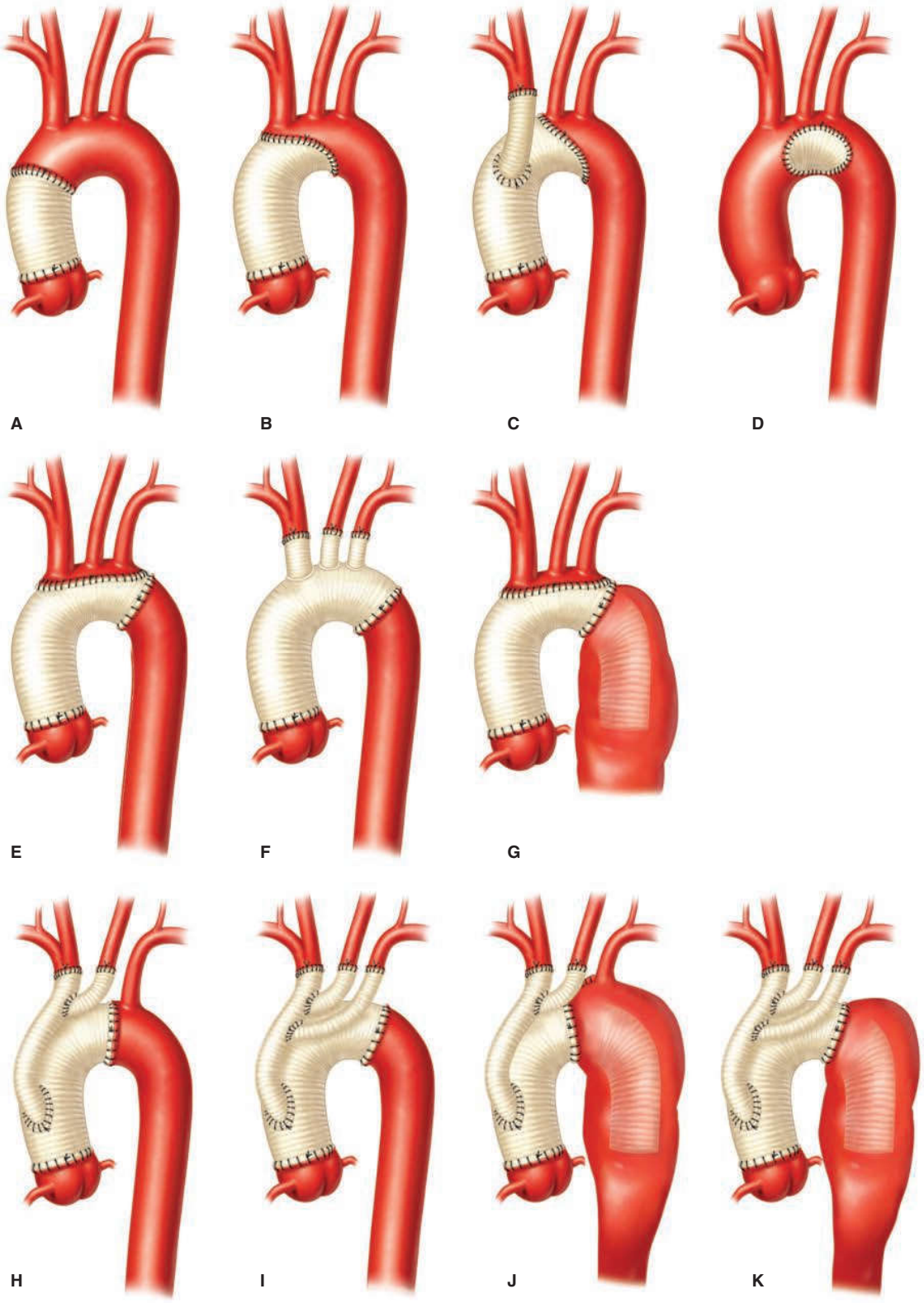


Figure 22-4. Illustrations of proximal aortic repairs in which the native aortic root is left intact. **A.** Graft replacement of the tubular portion of the ascending aorta with the aortic arch left intact. **B.** Hemiarch beveled graft replacement, in which the ascending aorta and a portion of the lesser curvature of the aortic arch are replaced. **C.** A modified arch with additional graft replacement of the innominate artery. **D.** Patch repair of the aortic arch. **E.** Traditional total arch replacement using an island approach to reattach the brachiocephalic vessels. **F.** The branched graft approach, which replaces the brachiocephalic vessels by following their original anatomic location. **G.** The elephant trunk approach with a concomitant island brachiocephalic artery reattachment. Contemporary Y-graft arch repairs include (**H**) the single Y-graft approach, (**I**) the double Y-graft approach, (**J**) the elephant trunk approach with a single Y-graft, and (**K**) the elephant trunk approach with a double Y-graft.

Table 22-2

Options for open surgical repair of proximal aortic aneurysms

- Options for treating aortic valve disease
  - Aortic valve annuloplasty (annular plication)
  - Aortic valve replacement (with mechanical or biologic prosthesis)
  - Aortic root replacement
    - Composite valve graft (with mechanical or biologic valve)
    - Aortic homograft
    - Stentless porcine root
    - Pulmonary autograft (Ross procedure)
    - Valve-sparing techniques
- Options for graft repair of the aortic aneurysm
  - Patch aortoplasty
  - Ascending replacement only
  - Beveled hemiarch replacement
  - Total arch replacement with reattachment of brachiocephalic branches (island technique)
    - Elephant trunk technique with island reattachment
  - Total arch replacement with bypass grafts to the brachiocephalic branches (including Y-graft approaches)
    - Elephant trunk technique with Y-graft approach
  - Hybrid aortic arch repairs (including "frozen elephant trunk technique")
- Perfusion options
  - Standard cardiopulmonary bypass
  - Profound hypothermic circulatory arrest without adjuncts
  - Hypothermic circulatory arrest with adjuncts
    - Retrograde cerebral perfusion
    - Antegrade cerebral perfusion
      - Balloon perfusion catheters
      - Right axillary artery cannulation
      - Innominate artery cannulation

arteries and brachiocephalic branches. The options for treating aortic valve disease, repairing aortic aneurysms, and maintaining perfusion during repair procedures each deserve detailed consideration (Table 22-2).

Aortic Valve Disease and Root Aneurysms Many patients undergoing proximal aortic operations have aortic valve disease that requires concomitant surgical correction. When such disease is present and the sinus segment is normal, separate repair or replacement of the aortic valve and graft replacement of the tubular segment of the ascending aorta are carried out. In such cases, mild to moderate valve regurgitation with annular dilatation can be addressed by plicating the annulus with mattress sutures placed below each commissure, thereby preserving the native valve. In patients with more severe valvular regurgitation or with valvular stenosis, the valve is replaced with a stented biologic or mechanical prosthesis; mechanical prostheses necessitate following a lifelong anticoagulation regimen. Separate replacement of the aortic valve and ascending aorta is not performed in patients with Marfan syndrome or Loeys-Dietz syndrome, because progressive dilatation of the remaining sinus segment eventually leads to complications that necessitate reoperation. Therefore, patients with Marfan syndrome, Loeys-Dietz syndrome, or annuloaortic ectasia require some form of aortic root replacement.[72]

In many cases, the aortic root is replaced with a mechanical or biologic graft that has both a valve and an aortic conduit. Currently, the following graft options are commercially available: composite valve grafts with a mechanical valve, which consist of a bileaflet mechanical valve attached to a polyester tube graft; composite valve grafts with a biological valve (available in Europe only at this point); aortic root homografts, which are harvested from cadavers and cryopreserved[73]; and stentless porcine aortic root grafts.[74,75] In the United States, because no biologic composite valve grafts are commercially available, another option for surgeons is to construct a bioprosthetic composite valve graft during the operation by suturing a stented tissue valve to a polyester tube graft.

Although select patients may be offered the Ross procedure—in which the patient's pulmonary artery root is excised and placed in the aortic position and then the right ventricular outflow tract is reconstructed by using a cryopreserved pulmonary homograft—this option is rarely used. This is largely because it is a technically demanding procedure, and there are concerns about the potential for autograft dilatation in patients with heritable conditions.[76]

An additional option is valve-sparing aortic root replacement, which has evolved substantially during the past decade.[77,78] The valve-sparing technique that is currently favored is called *aortic root reimplantation* and involves excising the aortic sinuses, attaching a prosthetic graft to the patient's annulus (Fig. 22-5), and resuspending the native aortic valve inside the graft. The superior hemodynamics of the native valve and the avoidance of anticoagulation are major advantages of the valve-sparing approach. Long-term results in carefully selected patients have been excellent.[79] Although the durability of this procedure in patients with Marfan syndrome has been satisfactory in some centers, it remains uncertain whether long-term durability can be reliably achieved with this approach.[78] Further, acceptable mid-term outcomes have been reported for patients with bicuspid aortic valve.[80] Patients who have structural leaflet deterioration or excessive annular dilatation are typically deemed unsuitable for valve-sparing repair.

Regardless of the type of conduit used, aortic root replacement requires reattaching the coronary arteries to openings in the graft. In the original procedure described by Bentall and De Bono,[81] this was accomplished by suturing the intact aortic wall surrounding each coronary artery to the openings in the graft. The aortic wall was then wrapped around the graft to establish hemostasis. However, this technique frequently produced leaks at the coronary reattachment sites that eventually led to pseudoaneurysm formation. Cabrol's modification, in which a separate, small tube graft is sutured to the coronary ostia and the main aortic graft, achieves tension-free coronary anastomoses, and reduces the risk of pseudoaneurysm formation.[82] Kouchoukos's button modification of the Bentall procedure is currently the most widely used technique.[83] The aneurysmal aorta is excised, and buttons of aortic wall are left surrounding both coronary arteries, which are then mobilized and sutured to the aortic graft (Fig. 22-6). The coronary suture lines may be reinforced with polytetrafluoroethylene felt or pericardium

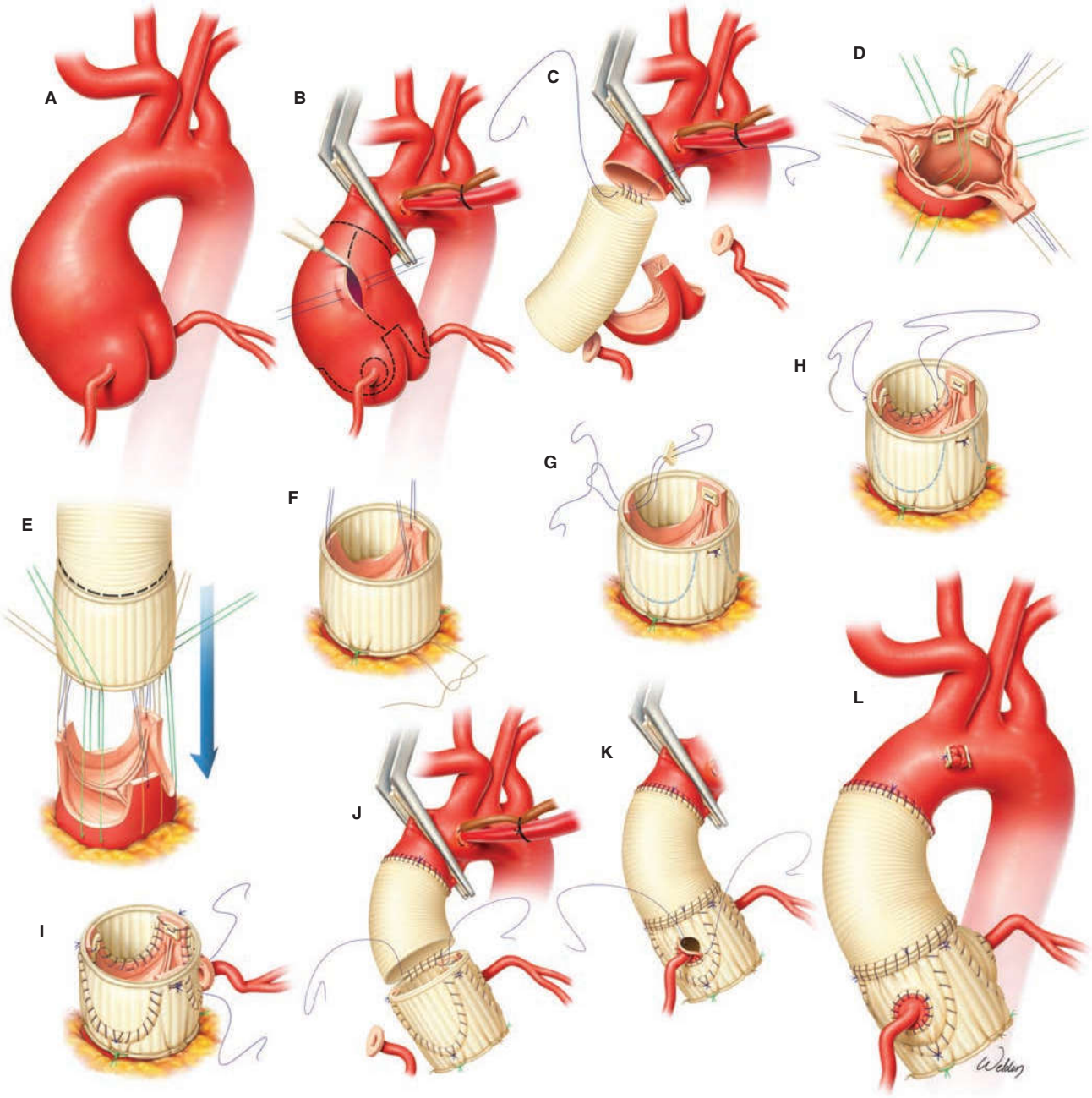


Figure 22-5. Illustration of our current valve-sparing procedure for replacing the aortic root and ascending aorta for treatment of (**A**) aortic root aneurysm. **B.** The ascending aorta is opened after cardiopulmonary bypass and cardioplegic arrest are established and the distal ascending aorta is clamped. The diseased aortic tissue (including the sinuses of Valsalva) is excised. Buttons of surrounding tissue are used to mobilize the origins of the coronary arteries. **C.** A synthetic graft is sewn to the distal ascending aorta with continuous suture. **D.** After the distal anastomosis is completed, six sutures reinforced with Teflon pledgets are placed in the plane immediately below the aortic valve annulus. **E.** The subannular sutures are placed through the base of a synthetic aortic root graft, which is then is parachuted down around the valve. **F.** After the root graft is cut to an appropriate length, the valve commissures and leaflets are positioned within the graft. The annular sutures are then tied. **G.** Each of the three commissures is then secured near the top of the graft. **H.** The supra-annular aortic tissue is sewn to the graft in continuous fashion. **I.** The button surrounding the origin of the left main coronary artery is sewn to an opening cut in the root graft. **J.** The two aortic grafts are sewn together with continuous suture. **K.** The button surrounding the origin of the right coronary artery is sewn to an opening cut in the root graft. **L.** The completed valve-sparing aortic root replacement and graft repair of the ascending aorta are shown. *(Used with permission of Baylor College of Medicine.)*

to enhance hemostasis. When the coronary arteries cannot be mobilized adequately because of extremely large aneurysms or scarring from previous surgery, the Cabrol technique or a related modification can be used. Another option, originally described by Zubiate and Kay,[84] is the construction of bypass grafts by using interposition saphenous vein or synthetic grafts.

Aortic Arch Aneurysms Several options are also available for handling aneurysms that extend into the transverse aortic arch

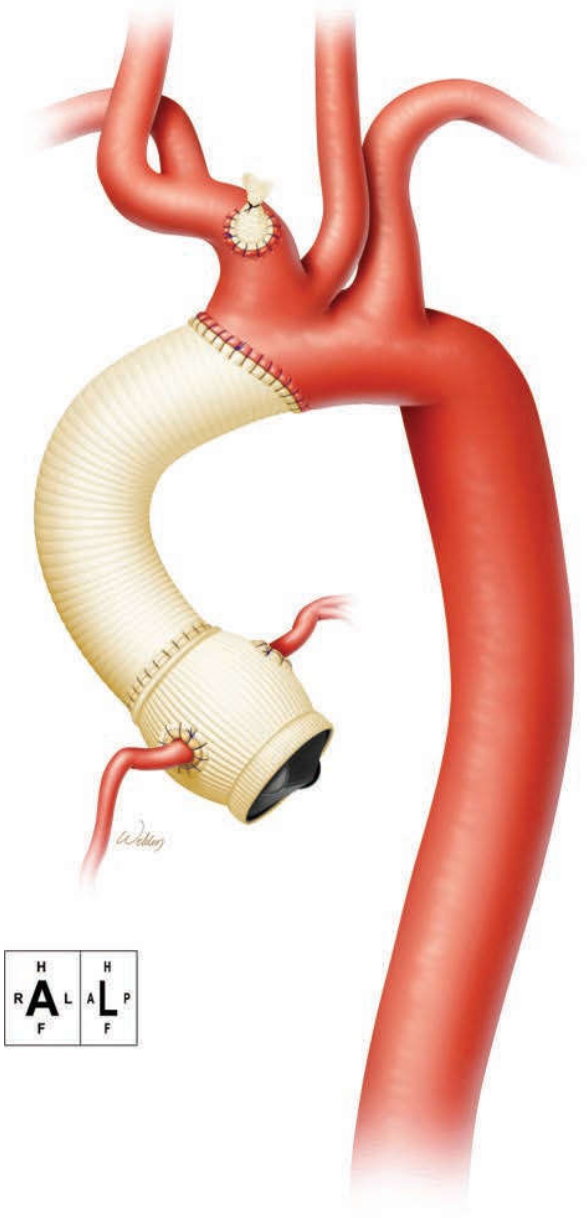


Figure 22-6. Illustration of the modified Bentall procedure for replacing the aortic root and ascending aorta. Commonly, the ascending aorta is replaced by a straight "tube" graft, and the aortic root, including the valve apparatus and the sinuses of Valsalva, is replaced by a mechanical composite valve graft with neosinuses to better mimic its native shape. The coronary arteries with buttons of surrounding aortic tissue have been mobilized and are being reattached to openings in the aortic graft. Shown at the proximal aspect of the innominate artery, the ligated remnant of an 8-mm graft was used to ease cannulation of the artery, which was used as inflow during cardiopulmonary bypass. *(Used with permission of Baylor College of Medicine.)*

(see Fig. 22-4C-K).[65] The surgical approach depends on the extent of involvement and the need for cardiac and cerebral protection. Saccular aneurysms that arise from the lesser curvature of the distal transverse arch and that encompass <50% of the aortic circumference can be treated by patch graft aortoplasty; such aneurysms are particularly well suited for hybrid repair with arch debranching followed by exclusion of the aneurysm with an endovascular graft.[85] For fusiform aneurysms, when the distal portion of the arch is a reasonable size, a single, beveled replacement of the lower curvature (hemiarch) is performed. More extensive arch aneurysms require total replacement involving a distal anastomosis to the proximal descending thoracic aorta and separate reattachment of the brachiocephalic branches. The brachiocephalic vessels can be reattached to one or more openings made in the graft, or if these vessels are aneurysmal, they can be replaced with separate, smaller grafts. Alternatively, a Y-graft approach[86,87] can be used to debranch the brachiocephalic vessels and move them forward, thereby permitting the distal anastomosis to be brought forward, which aids in hemostasis. When the aneurysm involves the entire arch and extends into the descending thoracic aorta, it is approached by using Borst's elephant trunk technique of staged total arch replacement.[88] The distal anastomosis is performed such that a portion of the graft is left suspended within the proximal descending thoracic aorta (Fig. 22-7). A collared graft can be used to accommodate any discrepancy in aortic diameter.[86] During a subsequent operation, the suspended "trunk" is used to facilitate repair of the remaining descending thoracic or thoracoabdominal aortic aneurysm by an endovascular technique or by open repair through a thoracotomy incision, depending on the extent of disease and other factors. The elephant trunk technique permits access to the distal portion of the graft during the second operation without the need for dissection around the distal transverse aortic arch; this reduces the risk of injuring the left recurrent laryngeal nerve, esophagus, and pulmonary artery if an open approach is used at the second stage. As described in the section on hybrid repair of arch aneurysms (see later), the elephant trunk can be completed by using a hybrid endovascular approach (Fig. 22-8) in certain settings. A newer technique that is currently under investigation involves using a graft comprising a conventional polyester proximal portion and a stent graft distal elephant trunk portion. This "frozen elephant trunk" technique can enable treatment of the entire aortic pathology during a single procedure or can facilitate a subsequent endovascular procedure (Fig. 22-9).[89-91]

Cardiopulmonary Bypass Perfusion Strategies Like the operations themselves, perfusion strategies used during proximal aortic surgery depend on the extent of the repair. Aneurysms that are isolated to the ascending segment can be replaced by using standard cardiopulmonary bypass and distal ascending aortic clamping. This provides constant perfusion of the brain and other vital organs during the repair. Aneurysms involving the transverse aortic arch, however, cannot be clamped during the repair, which necessitates the temporary withdrawal of cardiopulmonary bypass support; this is called *circulatory arrest.* To protect the brain and other vital organs during the circulatory arrest period, hypothermia must be initiated before pump flow is stopped. However, the deep levels of hypothermia (below 20°C) that have been traditionally used in open arch repair are not without risk, and pure hypothermic circulatory arrest continues to have substantial limitations. Importantly, although brief periods of total circulatory arrest generally are well tolerated at cold temperatures, as the duration of circulatory arrest increases, the well-recognized risks of brain injury and death increase dramatically. Additionally, deep levels of hypothermia are associated with coagulopathy.

Because of the inherent complexity of aortic arch repairs and their general tendency to require longer periods

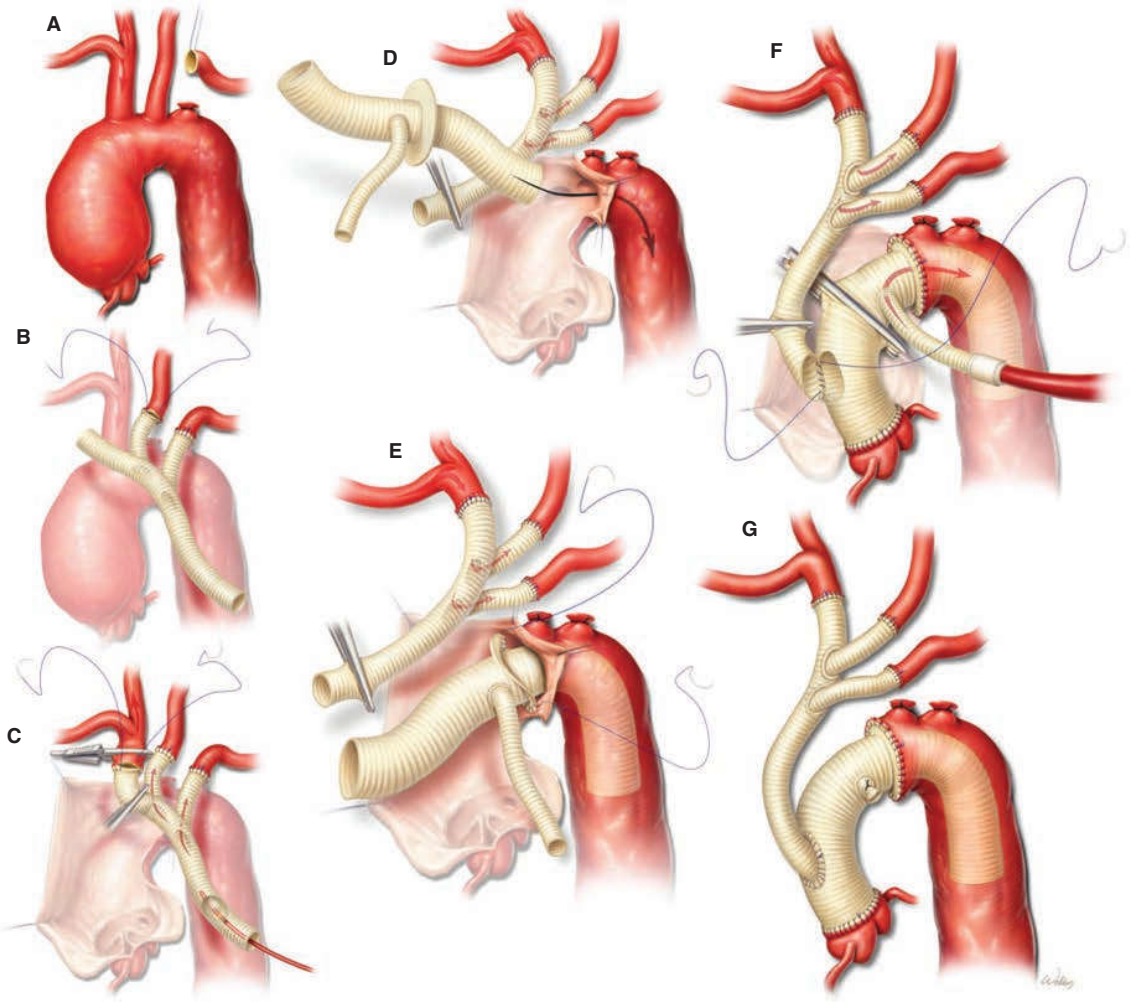


Figure 22-7. Illustration of a contemporary Y-graft approach to total arch replacement for aortic arch aneurysm. **A.** The proximal portions of the brachiocephalic arteries are exposed. **B.** The first two branches of the graft are sewn end-to-end to the transected left subclavian and left common carotid arteries. The proximal ends of the transected brachiocephalic arteries are ligated. **C.** A balloon-tipped perfusion cannula is placed inside the double Y-graft and used to deliver antegrade cerebral perfusion. After systemic circulatory arrest is initiated, the innominate artery is clamped, transected, and sewn to the distal end of the main graft. **D.** The proximal aspect of the Y-graft is clamped. This directs flow from the axillary artery to all three brachiocephalic arteries. The arch is then replaced with a collared elephant trunk graft. **E.** The distal anastomosis between the elephant trunk graft and the aorta is created between the innominate and left common carotid arteries. The collared graft accommodates any discrepancy in aortic diameter. **F.** The aortic graft is clamped, and a second limb from the arterial inflow tubing of the cardiopulmonary bypass circuit is used to deliver systemic perfusion through a side-branch of the arch graft while the proximal portion of the ascending aorta is replaced. Once the proximal aortic anastomosis is completed, the main trunk of the double Y-graft is cut to an appropriate length, and the beveled end is then sewn to an oval opening created in the right anterolateral aspect of the ascending aortic graft, which completes the repair (**G**). *(Modified with permission from LeMaire SA, Price MD, Parenti JL, et al. Early outcomes after aortic arch replacement by using the Y-graft technique,* Ann Thorac Surg. *2011 Mar;91(3):700-707.)*

of hypothermic circulatory arrest, two cerebral perfusion strategies—retrograde cerebral perfusion (RCP) and antegrade cerebral perfusion (ACP)—were developed to supplement this process by delivering cold, oxygenated blood to the brain and further reduce the risks associated with repair. Retrograde cerebral perfusion involves directing blood from the cardiopulmonary bypass circuit into the brain through the superior vena cava.[92] However, RCP is thought to be less beneficial than ACP,[93] and although it may be helpful in the retrograde flushing of air and debris from the arch, many centers have stopped using RCP.

In contrast, ACP delivers blood directly into the brachiocephalic arteries to maintain cerebral flow. Although its use was cumbersome in the past, contemporary ACP techniques (Fig. 22-10) have been simplified and commonly involve cannulating either the right axillary artery or the innominate artery and subsequent connection to the cardiopulmonary bypass circuit.[94-96] Often, a small section of graft is used as a conduit to ease cannulation, but there remains a small procedure-related risk of brachial plexus or vascular injury. Upon initiation, cold blood is delivered into the brain via the right common carotid artery and, if bilateral ACP is desired, the left common carotid artery. Note that, with the unilateral ACP technique, blood flow to the left side of the brain requires collateral circulation, ideally through an intact circle of Willis.

Methods to help determine the adequacy of unilateral ACP to deliver cerebral cross-circulation include preoperative imaging and intraoperative monitoring. A commonly used method

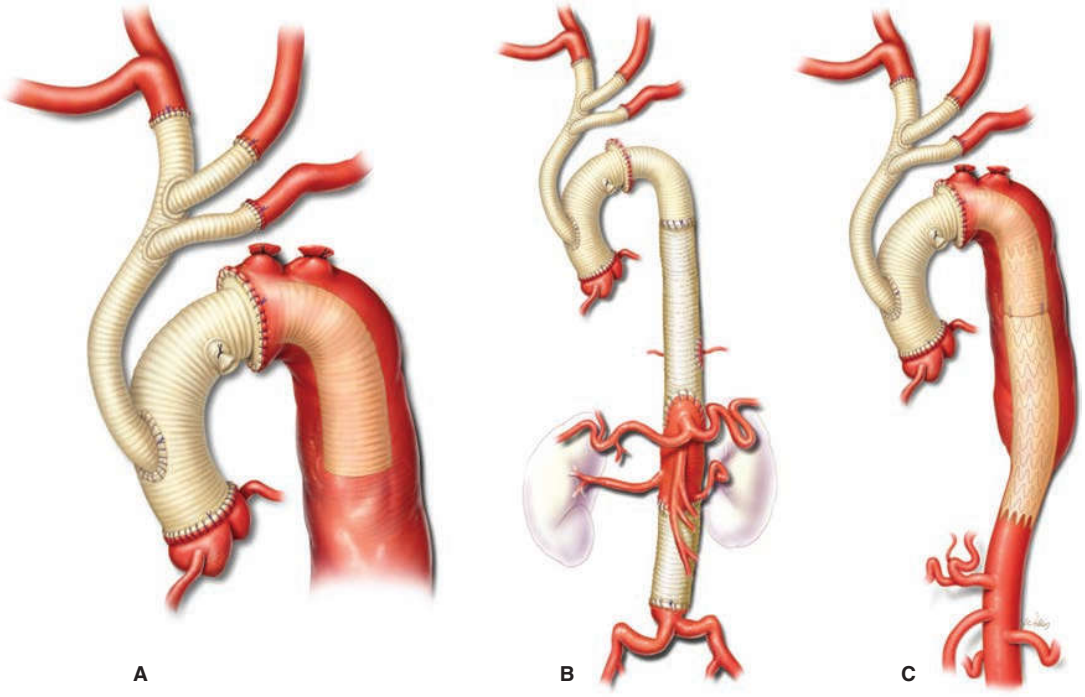


Figure 22-8. Illustration of Borst's elephant trunk technique using a contemporary Y-graft approach. **A.** Stage 1: The proximal repair includes replacing the ascending aorta and entire arch, with Y-graft reattachment of the brachiocephalic vessels. The distal anastomosis is facilitated by using a collared elephant trunk graft to accommodate the larger diameter of the distal aorta. A section of the graft is left suspended within the proximal descending thoracic aorta. **B.** Stage 2: The distal repair uses the floating "trunk" for the proximal anastomosis. **C.** An alternate "hybrid" approach may be used in patients with less extensive distal aortic disease. Endovascular stent grafts are placed within the elephant trunk to complete the repair. (*Used with permission of Baylor College of Medicine.)*

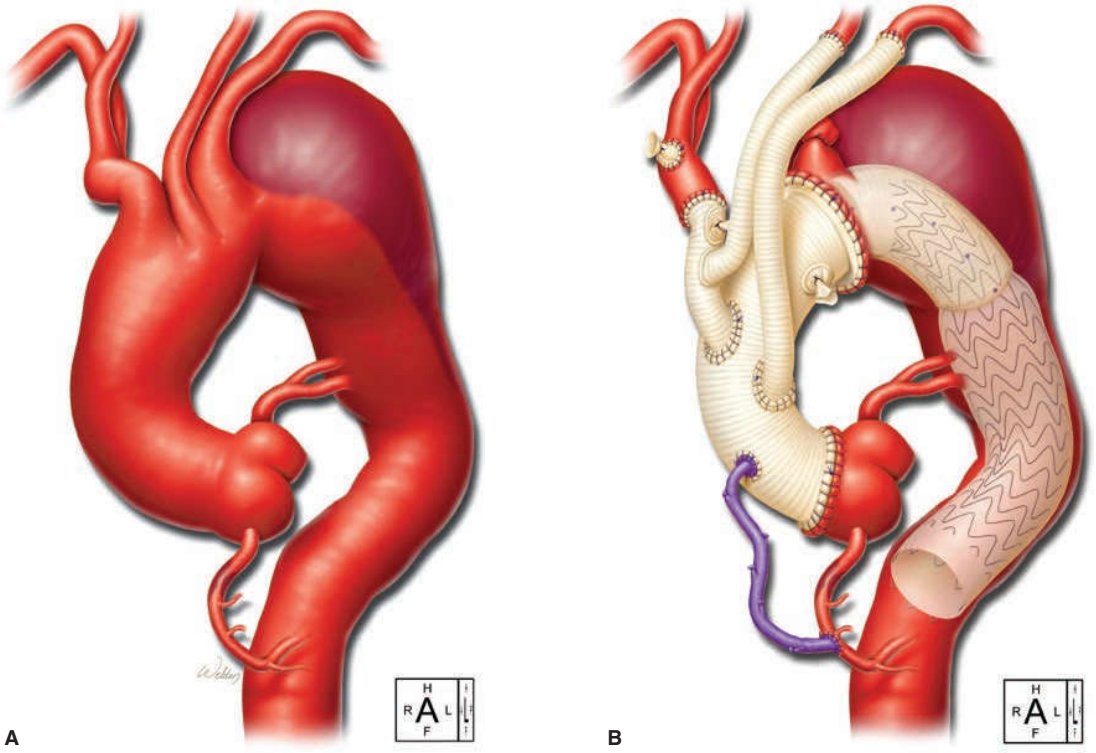


Figure 22-9. Illustration of a frozen elephant trunk repair, which is a hybrid approach to repair that combines open aortic replacement with endovascular aortic repair. **A.** Extensive aortic disease affects the proximal and distal aorta. **B.** Aortic repair is extended into the proximal portion of the descending thoracic aorta after the transverse aortic arch is fully replaced. (*Used with permission of Baylor College of Medicine.)*

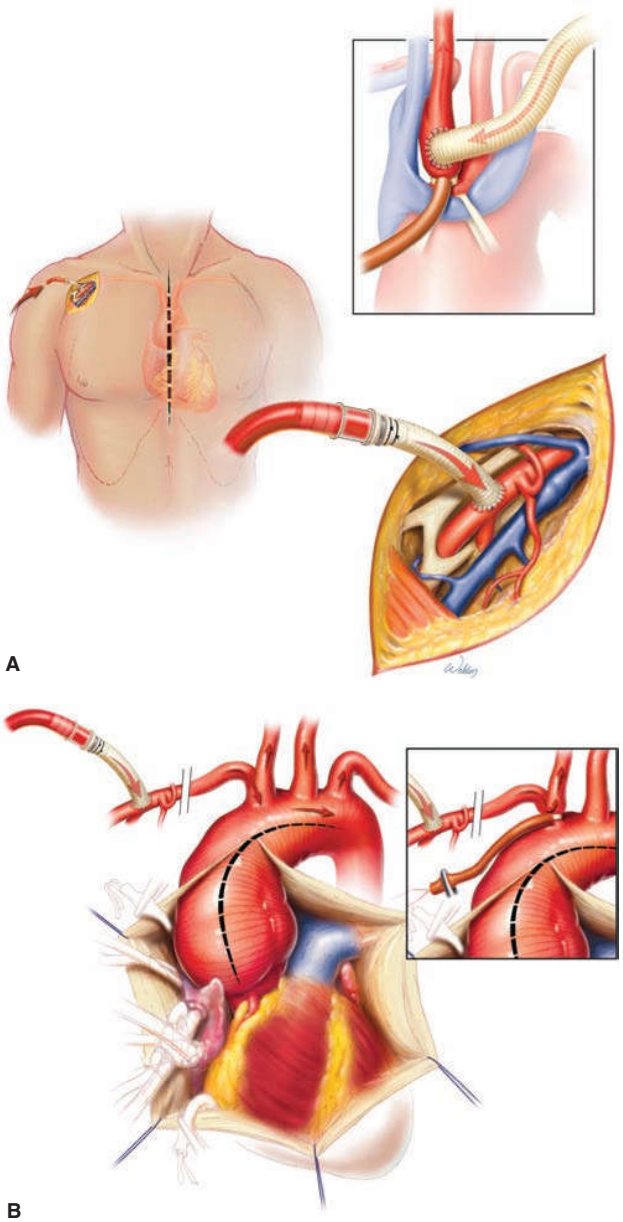


Figure 22-10. Illustration of a contemporary technique for delivering antegrade cerebral perfusion during aortic arch repair. **A.** A graft sewn to the right axillary artery or to the innominate artery (*inset*) is used to return oxygenated blood from the cardiopulmonary bypass circuit. **B.** After adequate hypothermia is established, the innominate artery is occluded with a tourniquet (*inset*) so that flow is diverted to the right common carotid artery, which maintains cerebral circulation. *(Used with permission from Baylor College of Medicine.)*

of intraoperative monitoring is brain near-infrared spectroscopy (NIRS), which measures cerebral oxygenation. If NIRS monitoring indicates inadequate perfusion, an additional perfusion catheter can be inserted into the left common carotid artery to provide blood flow to the left side of the brain.

Because ACP provides excellent brain protection, many surgeons now use more moderate levels of hypothermia (often between 22°C and 24°C) to decrease the risks associated with deep hypothermia.[97] However, some authors have raised the concern that reducing the degree of hypothermia increases the risk of ischemic complications involving the spinal cord, kidneys, and other organs that are ischemic (without the benefit of deep hypothermia) during the systemic circulatory arrest period.[98] Consequently, some groups supplement ACP with additional perfusion strategies that provide flow to the descending aorta during arch repair.[99,100]

Endovascular Repair Experience with purely endovascular treatment of proximal aortic disease remains limited and only investigational.[101] The unique anatomy of the aortic arch and the need for uninterrupted cerebral perfusion pose difficult challenges. There are reports of the use of "homemade" grafts to exclude arch aneurysms; however, these grafts are experimental at this time. For example, in 1999, Inoue and colleagues[102] reported placing a triple-branched stent graft in a patient with an aneurysm of the aortic arch. The three brachiocephalic branches were positioned by placing percutaneous wires in the right brachial, left carotid, and left brachial arteries. The patient underwent two subsequent procedures: surgical repair of a right brachial pseudoaneurysm and placement of a distal stent graft extension to control a major perigraft leak. Since then, efforts to employ endovascular techniques in the treatment of the proximal aorta have been essentially limited to the use of approved devices for off-label indications, such as the exclusion of pseudoaneurysms in the ascending aorta.

Hybrid Repair In June 1991, the Ukrainian surgeon Nikolay Volodos and his colleagues performed the first hybrid aortic arch repair[103,104]; 22 years later, Volodos reported that the patient was still alive.[105] Unlike purely endovascular approaches, hybrid repairs of the aortic arch have entered the mainstream clinical arena, although they remain controversial. Hybrid arch repairs involve some form of "debranching" of the brachiocephalic vessels (which are not unlike Y-graft approaches), followed by endovascular exclusion of some or all of the aortic arch (Fig. 22-11). Although this technique has many variants, they often involve sewing a branched graft to the proximal ascending aorta with the use of a partial aortic clamp. The branches of the graft are then sewn to the arch vessels. Once the arch is "debranched," the arch aneurysm can be excluded with an endograft. Commonly, a zone 0 approach (Fig. 22-12) is undertaken in which the proximal end of the endograft is secured within the ascending aorta. Other hybrid approaches aim to extend repair into the distal arch and descending thoracic aorta (see the following section). The arguments for using a hybrid approach to treat aortic arch aneurysm include the potential to avoid using cardiopulmonary bypass, circulatory arrest, and cardiac ischemia.[59,60]

It is not yet clear whether hybrid repairs are as durable as traditional ones because little mid- or long-term data have been published, and there are very few comparative studies.[65] Procedure-related risks include the risk of embolization and stroke due to wire and device manipulation within the aortic arch (this risk appears to be greatest in zone 0 repairs[106]), retrograde acute aortic dissection,[107] type I endoleak,[108] and paraplegia.[27] Because iatrogenic retrograde dissection of the ascending aorta is a particularly lethal complication, special considerations, including careful blood pressure management and wire manipulation, are recommended to avoid this problem in patients who are undergoing hybrid arch zone 0 stent deployment.[109] Notably, patients with an ascending aortic diameter greater than 4.2 cm may be more susceptible to retrograde dissection. In an effort to

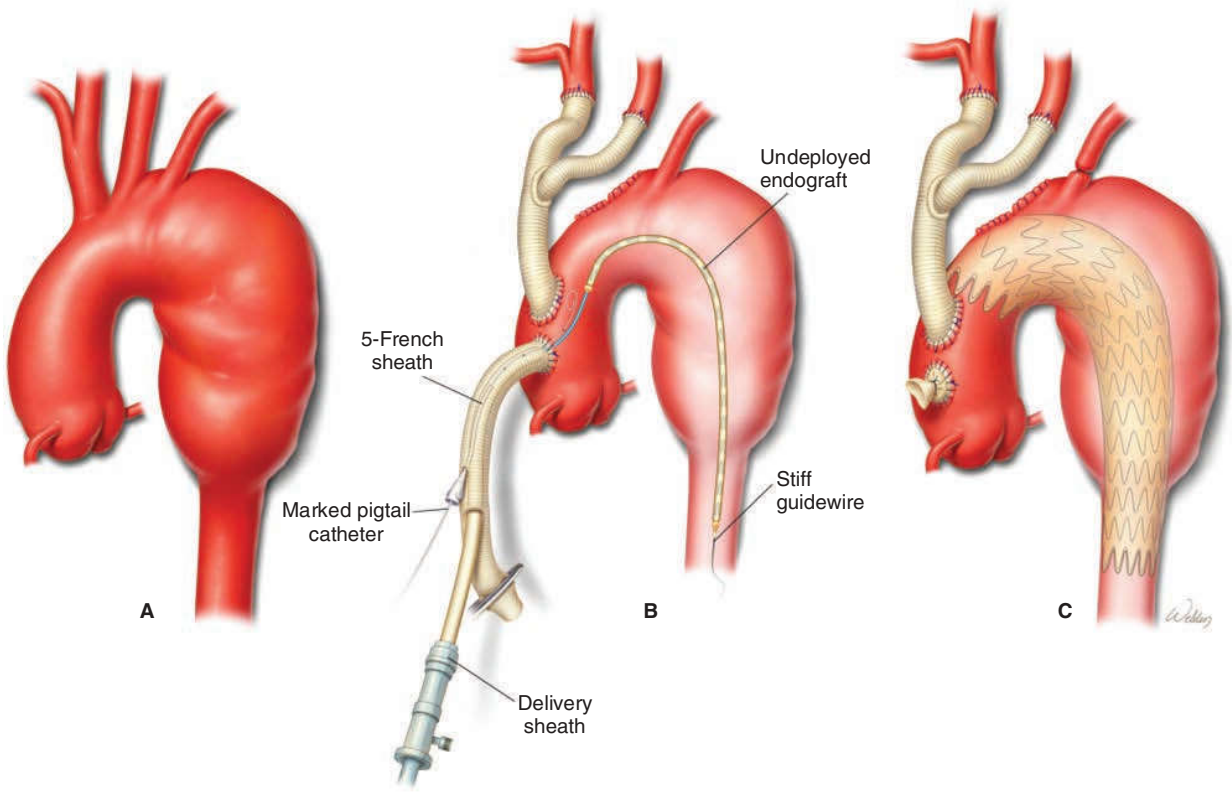


Figure 22-11. Illustration of a "Zone 0" hybrid arch repair. **A.** A distal arch aneurysm is shown that extends into the proximal aspect of the descending thoracic aorta. **B.** The brachiocephalic vessels are debranched onto a Y-graft, and a separate graft is used as a conduit for antegrade endovascular deployment of the stent graft. **C.** In the completed repair, the proximal landing zone of the endograft is within zone 0. *(Used with permission from Baylor College of Medicine.)*

reduce the risk of iatrogenic dissection, some centers have begun to replace a small section of the ascending aorta with a standard polyester graft such that the endograft's proximal landing zone comprises prosthetic material rather than native aortic tissue.[107]

Distal Thoracic Aortic Aneurysms

Open Repair In patients with descending thoracic or thoracoabdominal aortic aneurysms, several aspects of treatment—including preoperative risk assessment, anesthetic management, choice of incision, and use of protective adjuncts—are dictated by the overall extent of aortic involvement. By definition, descending thoracic aortic aneurysms involve the portion of the aorta between the left subclavian artery and the diaphragm. Thoracoabdominal aneurysms can involve the entire thoracoabdominal aorta, from the origin of the left subclavian artery to the aortic bifurcation. Surgical repair of thoracoabdominal aortic aneurysms is categorized by the extent of aortic replacement according to the Crawford classification scheme (Fig. 22-13). Extent I thoracoabdominal aortic aneurysm repairs involve most of the descending thoracic aorta, usually beginning near the left subclavian artery, and extend down into the suprarenal abdominal aorta. Extent II repairs also begin near the left subclavian artery but extend distally into the infrarenal abdominal aorta, and they often reach the aortic bifurcation. Extent III repairs extend from the lower descending thoracic aorta (below the sixth rib) and into the abdomen. Extent IV repairs begin at the diaphragmatic hiatus and often involve the entire abdominal aorta.

Descending thoracic aortic aneurysms not amenable to endovascular therapy are currently repaired through a left thoracotomy. In patients with thoracoabdominal aortic aneurysm, the thoracotomy is extended across the costal margin and into the abdomen.[110] Using a double-lumen endobronchial tube allows selective ventilation of the right lung and deflation of the left lung. Transperitoneal exposure of the thoracoabdominal aorta is achieved by performing medial visceral rotation and circumferential division of the diaphragm. During a period of aortic clamping, the diseased segment is replaced with a polyester tube graft. Important branch arteries—including intercostal arteries and the celiac, superior mesenteric, and renal arteries—are reattached to openings made in the side of the graft. In patients with Marfan syndrome and other heritable conditions, separate (8- and 10-mm) grafts to the visceral branches are often used to prevent subsequent "patch aneurysms" that can develop in residual aortic tissue.[111] Visceral and renal artery occlusive disease is commonly encountered during aneurysm repair; options for correcting branch-vessel stenosis include endarterectomy, direct arterial stenting, and bypass grafting.

Clamping the descending thoracic aorta causes ischemia of the spinal cord and abdominal viscera. Clinically significant manifestations of hepatic, pancreatic, and bowel ischemia are relatively uncommon. However, both acute renal failure and spinal cord injury resulting in paraplegia or paraparesis remain major causes of morbidity and mortality after these operations. Therefore, several aspects of the operation are devoted to minimizing spinal and renal ischemia (Table 22-3). Our multimodal approach to spinal cord protection includes expeditious repair to minimize aortic clamping time, moderate systemic heparinization (1.0 mg/kg) to prevent small-vessel

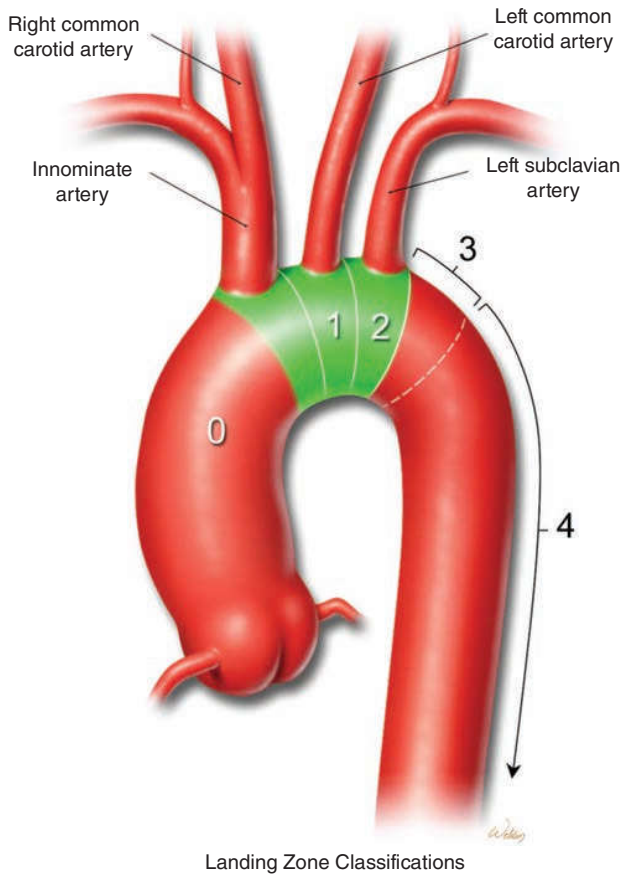


Figure 22-12. Illustration of the Criado landing zones, which are used to describe aortic anatomy during thoracic endovascular repair. The arch is the short segment that includes the origins of the three brachiocephalic arteries—the innominate artery, the left common carotid artery, and the left subclavian artery. Zone 0 includes the ascending aorta and the origin of the innominate artery. Zone 1 includes the origin of the left common carotid artery. Zone 2 includes the left subclavian artery origin. Zone 3 is a short section of the aorta that comprises the 2 cm immediately distal to the origin of the left subclavian artery, and zone 4 begins where zone 3 ends. *(Used with permission from Baylor College of Medicine.)*

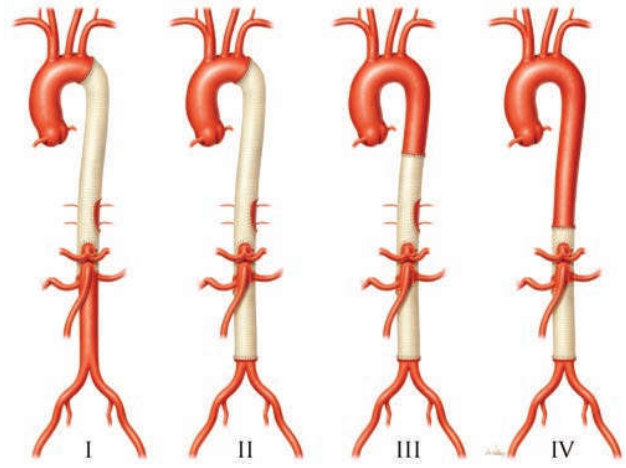


Figure 22-13. Illustration of the Crawford classification of thoracoabdominal aortic aneurysm repair, based on the extent of aortic replacement. *(Used with permission from Baylor College of Medicine.)*

Table 22-3

Current strategy for spinal cord and visceral protection during repair of distal thoracic aortic aneurysms

All extents
- Permissive mild hypothermia (32°C–34°C, nasopharyngeal)
- Moderate heparinization (1 mg/kg)
- Aggressive reattachment of segmental arteries, especially between T8 and L1
- Sequential aortic clamping when possible
- Perfusion of renal arteries with 4°C crystalloid solution when possible

Crawford extent I and II thoracoabdominal repairs
- Cerebrospinal fluid drainage
- Left heart bypass during proximal anastomosis
- Selective perfusion of celiac axis and superior mesenteric artery during intercostal and visceral anastomoses

thrombosis, mild permissive hypothermia (32°C to 34°C [89.6°F to 93.2°F] nasopharyngeal temperature), and reattachment of segmental intercostal and lumbar arteries. As the aorta is replaced from proximal to distal, the aortic clamp is moved sequentially to lower positions along the graft to restore perfusion to newly reattached branch vessels. During extensive thoracoabdominal aortic repairs (i.e., Crawford extent I and II repairs), cerebrospinal fluid drainage is used to improve spinal perfusion by reducing cerebrospinal fluid pressure. Because the benefits of this adjunct have been confirmed in a randomized clinical trial,[112] its use is recommended in current guidelines (Class I, Level B recommendation).[44] During cerebral spinal fluid drainage, the cerebral spinal fluid pressure is closely monitored, and the amount of fluid that is removed is carefully limited to avoid the devastating complication of intracranial hemorrhage.[113] Motor evoked potentials are used by some groups to monitor the spinal cord throughout the operation.[114,115] Left heart bypass, which provides perfusion of the distal aorta and its branches during the clamping period, is also used during extensive thoracoabdominal aortic repairs.[116-118] Because left heart bypass unloads the heart, it is also useful in patients with poor cardiac reserve. Balloon perfusion cannulas connected to the left heart bypass circuit can be used to deliver blood directly to the celiac axis and superior mesenteric artery during their reattachment. The potential benefits of reducing hepatic and bowel ischemia include reduced risks of postoperative coagulopathy and bacterial translocation, respectively. Whenever possible, renal protection is achieved by perfusing the kidneys with cold (4°C [39.2°F]) crystalloid. In a randomized clinical trial, reduced kidney temperature was found to be associated with renal protection, and the use of cold crystalloid independently predicted preserved renal function.[119]

Hypothermic circulatory arrest can also be used during descending thoracic or thoracoabdominal aortic repairs.[120] At our center, the primary indication for this approach is the inability to clamp the aorta because of rupture, extremely large aneurysm size, or extension of the aneurysm into the distal transverse aortic arch, or because a prior endovascular repair hinders clamping.[67]

As discussed previously, complete repair of extensive aneurysm involving the ascending aorta, transverse arch, and descending thoracic aorta generally requires staged open

operations or a hybrid approach. In such procedures, when the descending or thoracoabdominal component is symptomatic (e.g., causes back pain or has ruptured) or is disproportionately large (compared with the ascending aorta), the distal segment is treated during the initial operation, and repair of the ascending aorta and transverse aortic arch is performed as a second procedure. A reversed elephant trunk repair, in which a portion of the proximal end of the aortic graft is inverted down into the lumen, can be performed during the first operation; this technique facilitates the second-stage repair of the ascending aorta and transverse aortic arch (Fig. 22-14).[121]

Although spinal cord ischemia and renal failure receive the most attention, several other complications warrant consideration. The most common complication of extensive repairs is pulmonary dysfunction. With aneurysms adjacent to the left subclavian artery, the vagus and left recurrent laryngeal nerves are often adherent to the aortic wall and thus are susceptible to injury. Vocal cord paralysis should be suspected in patients who have postoperative hoarseness, and the presence of nerve damage should be confirmed by endoscopic examination. Vocal cord paralysis can be treated effectively by direct cord medialization (type 1 thyroplasty).[122] Injury to the esophagus during

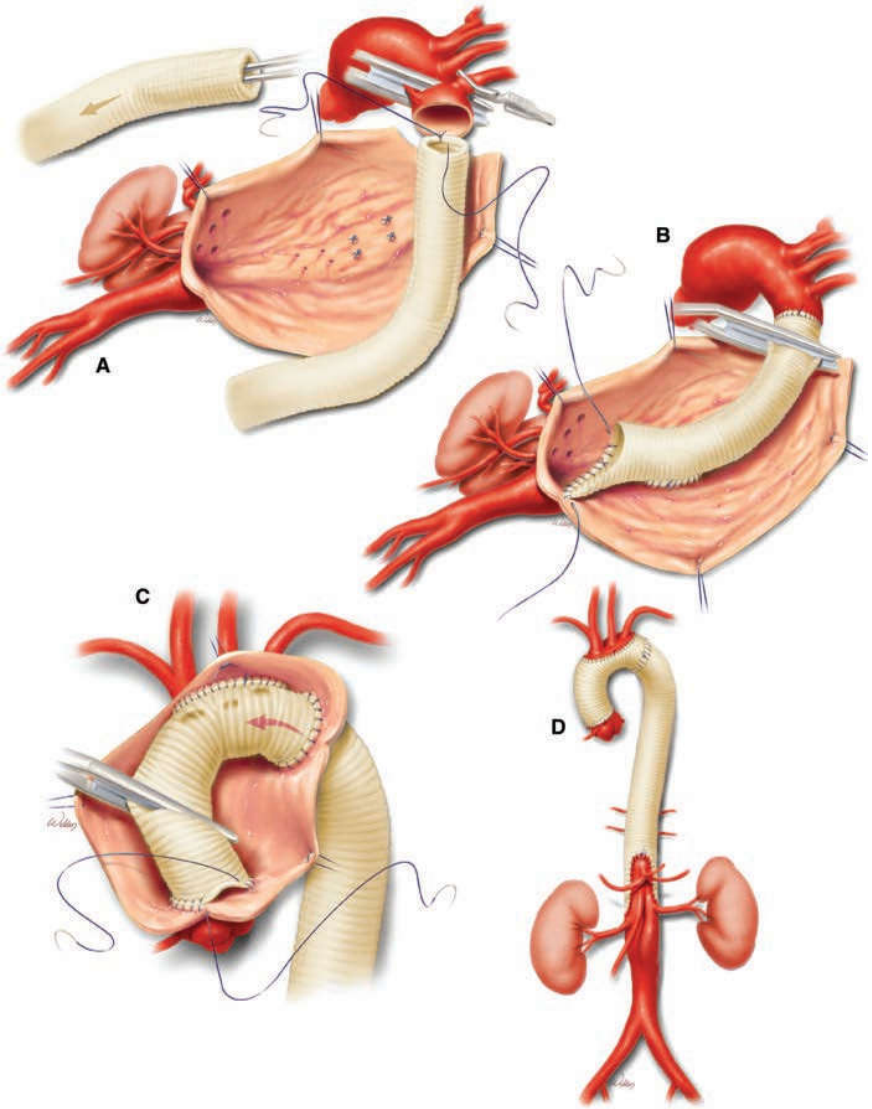


Figure 22-14. Illustration of the reversed elephant trunk technique using a traditional "island" approach to total aortic arch replacement. **A.** Stage 1: The distal aorta is repaired through a left thoracoabdominal approach. The aneurysm is opened after the aorta is clamped between the left common carotid artery and the left subclavian artery, which is also clamped. Before the proximal anastomosis is performed, the end of the graft is partly invaginated to leave a "trunk" for the subsequent repair. Proximal intercostal arteries are oversewn. **B.** After the proximal suture line is completed, the clamps are repositioned to restore blood flow to the left subclavian artery. The repair is completed by reattaching patent intercostal arteries to an opening in the side of the graft and creating a beveled distal anastomosis at the level of the visceral branches. **C.** Stage 2: The proximal aorta is repaired through a median sternotomy. The aortic arch is opened under hypothermic circulatory arrest. The "trunk" is pulled out and used to replace the aortic arch and ascending aorta. This eliminates the need for a new distal anastomosis and simplifies the procedure. Circulatory arrest and operative time, along with their attendant risks, are reduced. **D.** The completed two-stage repair of the entire thoracic aorta. *(Modified with permission from Coselli JS, LeMaire SA, Carter SA, et al: The reversed elephant trunk technique used for treatment of complex aneurysms of the entire thoracic aorta,* Ann Thorac Surg. *2005 Dec;80(6):2166-2172.)*

the proximal anastomosis can have catastrophic consequences. Carefully separating the proximal descending thoracic aorta from the underlying esophagus before performing the proximal anastomosis minimizes the risk of a secondary aortoesophageal fistula. In patients who have previously undergone coronary artery bypass with a left internal thoracic artery graft, clamping proximal to the left subclavian artery can precipitate severe myocardial ischemia and cardiac arrest. When the need to clamp at this location is anticipated in these patients, a left common carotid-to-subclavian bypass is performed to prevent cardiac complications (Fig. 22-15).[123]

Endovascular Repair

Descending Thoracic Aortic Aneurysms Stent graft repair has become the standard treatment for patients with descending thoracic aortic aneurysm.[55,56,124] Although aortic repair with a self-fixing endoprosthesis was reported by Volodos[103,104] in the mid 1980s, it was the report by Parodi and associates[125] of using endovascular stent grafting to repair abdominal aortic aneurysm that launched widespread interest in developing this approach. Only 3 years after this seminal report was published, Dake and colleagues[126] reported performing endovascular descending thoracic aortic repair with "homemade" stent grafts in 13 patients.

Guidelines for the use of endovascular repair in thoracic aortic disease have been published,[44] and reporting standards to uniformly describe the endovascular repair process have been established.[127] Although endografting was initially approved to treat degenerative descending thoracic aortic aneurysm, newer devices have been approved for use in treating various descending thoracic aortic pathologies, including blunt aortic injury, penetrating aortic ulcer (see following section), coarctation, and dissection. Although the use of stent grafts in cases of aortic infection is not ideal, patients with a fistula or mycotic aneurysm are sometimes treated with endovascular devices as a bridge to open repair.

In elderly patients with severe comorbidity and patients who have undergone previous complex thoracic aortic procedures, endovascular repair is a particularly attractive alternative to standard open surgical procedures.[128] Patients who undergo endovascular repair tend to have a lower incidence of intraoperative complications, a shorter length of stay, and a higher likelihood of being discharged to home than those who undergo open repair.[129] As mentioned previously, appropriate patient selection depends on specific measurements taken from preoperative CT angiograms.

To protect patients against spinal cord ischemia during endovascular repair of the descending thoracic aorta, the most important maneuver is to keep the mean arterial perfusion pressure between 90 and 110 mmHg after the endograft is deployed. In patients who have had previous open or endovascular abdominal aortic aneurysm repair, cerebrospinal fluid drainage is recommended.[130] The first step in the repair procedure is to obtain appropriate vascular access for the insertion of the thoracic stent graft. If the femoral artery will not accommodate the necessary

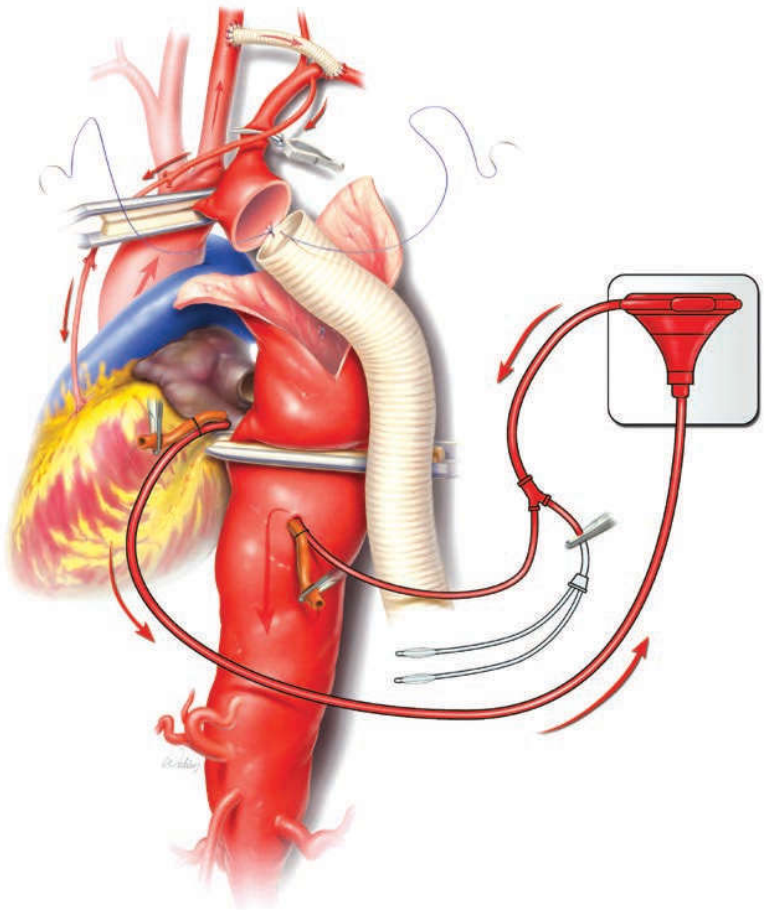


Figure 22-15. Illustration of a thoracoabdominal aortic aneurysm repair in a patient with a patent left internal thoracic artery-to-left anterior descending coronary artery graft. The proximal anastomosis is being performed while the aorta is clamped between the left common carotid and subclavian arteries. Myocardial perfusion is maintained through the carotid-subclavian bypass graft. *(Modifed with permission from Jones MM, Akay M, Murariu D, et al: Safe aortic arch clamping in patients with patent internal thoracic artery grafts.* Ann Thorac Surg. *2010 Apr;89(4):e31-e32.)*

sheath, then an iliac artery is exposed. If necessary, a graft can be sewn to the iliac artery in an end-to-side fashion to facilitate the deployment of the endograft. After 5,000 to 10,000 units of heparin are administered, a guidewire and the delivery sheath are typically inserted into the access artery under fluoroscopic guidance; depending on which endovascular device is used, the stent graft can be advanced through a sheath or with no sheath. The endograft is then advanced into the aorta and suitably positioned. Note that the best view of the distal arch and descending thoracic aorta is usually in the left anterior oblique position at an angle of approximately 40° to 50°. The device is then deployed, and the proximal and distal ends can be ballooned for better apposition of the stent graft to the aortic wall. An aortogram is then performed to rule out any endoleak, and protamine is administered. As an alternative to aortography, intravascular ultrasonography (IVUS) can be used to identify the proximal and distal landing zones, and the entire procedure can be performed with minimal or no contrast.

Although it is not uncommon to cover the left subclavian artery with the endograft to lengthen the proximal landing zone,[131] findings suggest that the risk of spinal cord complications is heightened when the subclavian artery is covered and not revascularized, presumably because of a loss of collateral circulation to the spinal cord.[132] To prevent this complication, a carotid-to-subclavian bypass can be easily constructed to maintain vertebral artery blood flow and minimize neurologic injury (Fig. 22-16).[133,134] In addition, recent studies suggest that revascularization of the left subclavian artery is associated with lower stroke risk in patients in whom an endograft was deployed in Zone 2 and covered the left subclavian artery.[135] In addition, new generations of stent grafts are being designed with side branches that can be placed within the left subclavian artery. This feature is particularly attractive if the proximal neck is short or if the patient has a patent left internal thoracic artery-to-left anterior descending coronary artery bypass. Indications for left subclavian artery revascularization include previous coronary artery bypass with patent internal thoracic artery, dominant left vertebral artery, aneurysm arising from the left subclavian artery, left arm arterio-venous fistula, and coverage of a long segment of the descending thoracic aorta.

Elephant Trunk Completion In select patients, elephant trunk completion repairs can be done with an endovascular approach (see Fig. 22-8C), rather than by the traditional open operation through a thoracotomy.[136] Recall that an elephant trunk is used when an aortic aneurysm extends from the distal arch to the descending thoracic aorta. An endograft can be deployed at the time of elephant trunk construction or during a separate, subsequent procedure.[97,107,137] When the stent is deployed in a retrograde manner during a second-stage procedure, the procedure is facilitated by placing radiopaque markers at the end of the elephant trunk during the first-stage procedure. This allows the distal end of the trunk to be identified via fluoroscopy. A guidewire can then be manipulated into the trunk and advanced into the ascending aorta to stabilize it during stent deployment. Note that advancing a wire in retrograde fashion from the femoral artery into the elephant trunk can be challenging. Occasionally, the wire must be advanced in an antegrade fashion from a brachial artery. The frozen elephant trunk technique—in which a short stent graft is delivered antegrade inside the trunk—can be used to perform the entire repair in one stage or to facilitate the second stage.[89,90]

Thoracoabdominal Aortic Aneurysms Although endovascular thoracoabdominal aortic aneurysm repair remains experimental, it has been shown to be feasible in a handful of specialized centers. Endovascular thoracoabdominal aortic aneurysm repairs are quite complex, because at least one of the visceral arteries is incorporated into the repair. The number of visceral branches that need to be addressed varies with the extent of aortic coverage.[138] The types of stent grafts used include fenestrated grafts, reinforced fenestrated grafts, branched or cuffed grafts, modular combinations of grafts, and multilayer stents.[139] Graft fenestrations and branch vessels are typically aligned by using inflatable angioplasty balloons. Procedure time is not insignificant, nor is the amount of contrast medium required to obtain the highly detailed images needed to plan these procedures. In addition, some of the stent grafts used in endovascular thoracoabdominal aortic aneurysm repair are custom-made in advance and thus may take several weeks to obtain; therefore, their use is limited to cases of elective repair.[107] In efforts to hasten repair and utilize off-the-shelf devices, parallel graft approaches, which use a combination of large- and small-diameter stents, have been reported.[140] And, although some centers now propose distal coverage of the celiac axis[141] for extent I thoracoabdominal aortic aneurysm repairs, this potentially risky approach is not widely used.

It should be noted that, like open thoracoabdominal aortic aneurysm repair, endovascular repair carries risks of paraplegia, renal failure, stroke, and death, despite the apparent benefits of its being a less invasive procedure. Notably, reports from centers experienced in endovascular thoracoabdominal aortic repair primarily describe limited extent IV repairs.[64] Although the technology is progressing rapidly, at present endovascular thoracoabdominal aortic aneurysm repair should be considered investigational.

Hybrid Repair Extensive hybrid thoracoabdominal aortic aneurysm repair[142,143] can be a life-saving option in patients at high surgical risk, such as those who have limited physiologic reserve, are of advanced age, or have significant comorbidities. Hybrid procedures use open surgical techniques to reroute blood supply to the visceral arteries so that their aortic origins can be covered by stent grafts without causing visceral ischemia (Fig. 22-17). Endovascular methods are then used (either as part of the same procedure or at a later stage) to repair the aortic aneurysm, often with simple tube stent grafts; such devices are more readily available than the customized, modular stent grafts deployed in strictly endovascular repairs. Overall, results for hybrid thoracoabdominal aortic aneurysm repair have been somewhat disappointing.[144] However, a handful of centers report acceptable outcomes in high-risk patients, particularly when a staged hybrid approach is used.[145]

Postoperative Considerations

Open Procedures Aortic anastomoses are often extremely fragile during the early postoperative period. Even brief episodes of postoperative hypertension can disrupt suture lines and precipitate severe bleeding or pseudoaneurysm formation. Therefore, during the initial 24 to 48 hours, meticulous blood pressure control is maintained to protect the integrity of the anastomoses. Generally, we liberally use IV vasoactive agents to keep the mean arterial blood pressure between 80 and 90 mmHg. In patients with extremely friable aortic tissue, such as those with Marfan syndrome, we lower the target range to 70 to 80 mmHg. It is a delicate balancing act because one must be

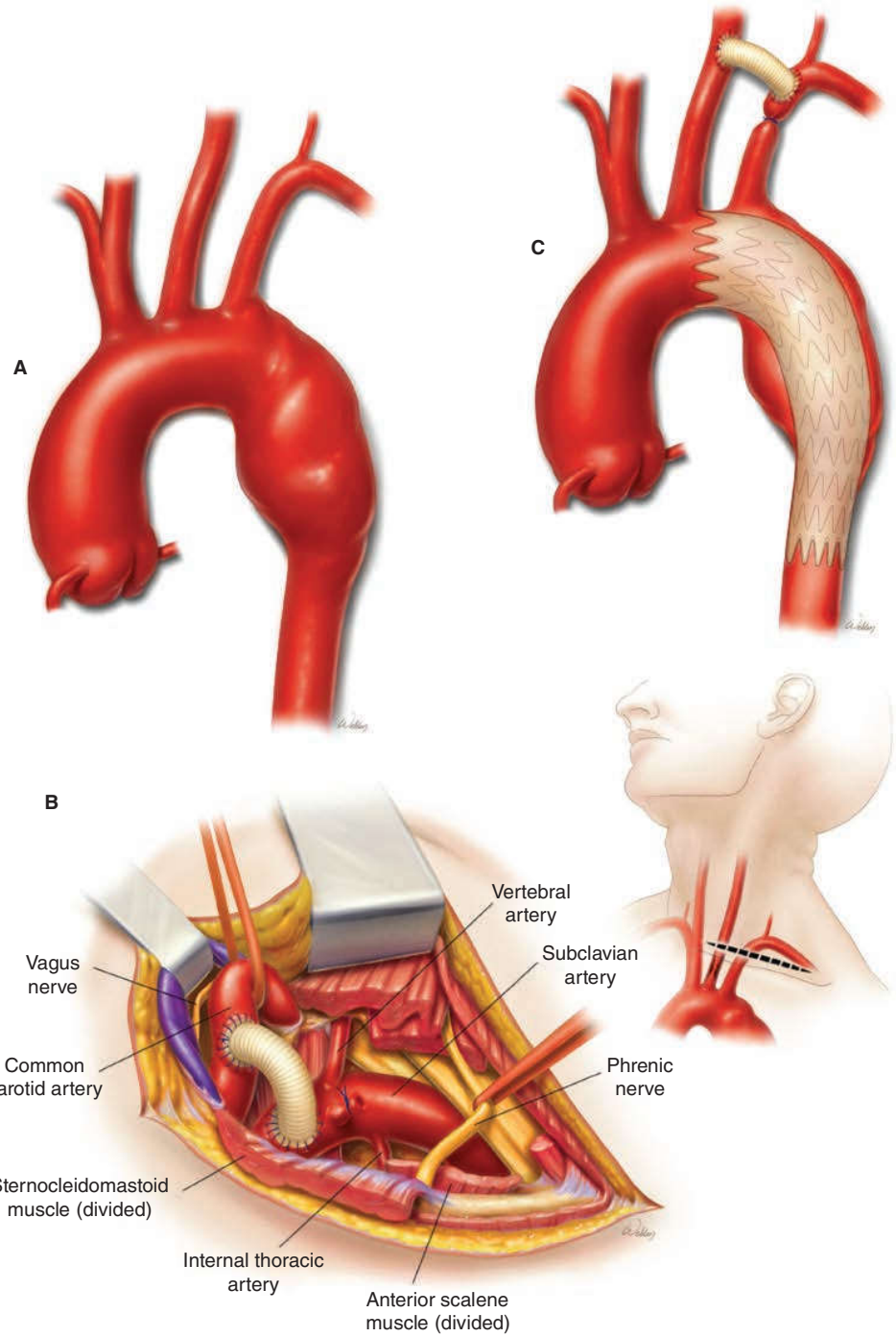


Figure 22-16. Illustration of a "Zone 2" hybrid repair of the proximal descending thoracic aorta. **A.** The preoperative representation of the aneurysm shows that establishing a 2-cm proximal landing zone for a stent graft will require covering the origin of the left subclavian artery. **B.** Through a supraclavicular approach, a bypass from the left common carotid artery to the left subclavian artery is performed to reroute circulation and create a landing zone for the stent graft. After the bypass is completed, the left subclavian artery is ligated proximal to the graft. **C.** In the completed hybrid repair, the aneurysm has been excluded successfully by a stent graft that covers the origin of the left subclavian artery, and the proximal landing zone of the endograft is within zone 2. Importantly, blood flow to the left vertebral artery and arm is preserved by the bypass graft. *(Reproduced with permission from Bozinovski J, LeMaire SA, Weldon SA: Hybrid Repairs of the Distal Aortic Arch and Proximal Descending Thoracic Aorta,* Oper Tech Thorac Cardiovasc Surg *2007;12(3):167-177.)*

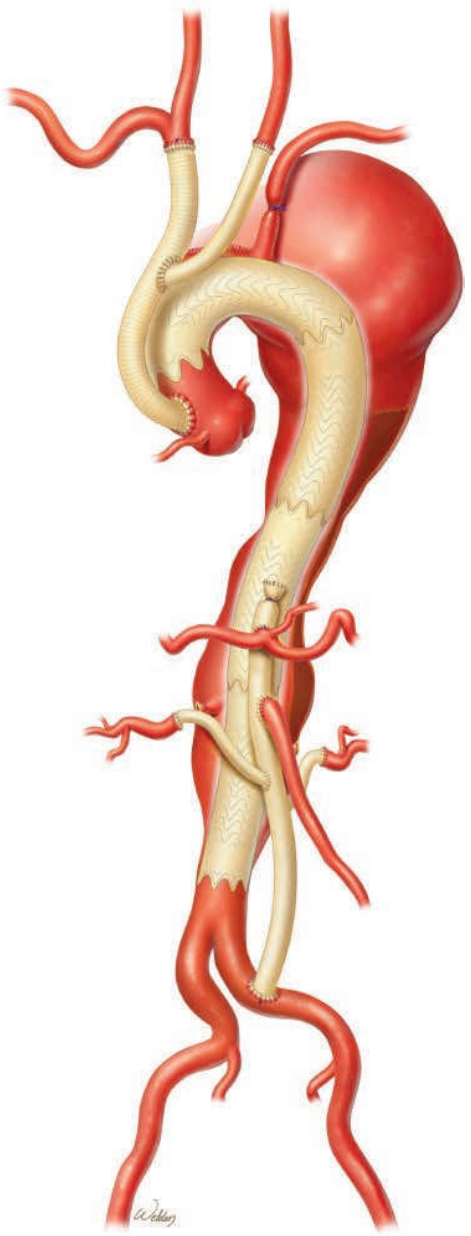


Figure 22-17. Illustration of a "Zone 0" hybrid approach—which combines open and endovascular techniques—for repair of an extensive aortic aneurysm. Debranching the arch and thoracoabdominal segments allows the use of a series of endovascular stent grafts to exclude the entire aneurysm. Note that the arterial inflow for the debranched visceral arteries comes from the left common iliac artery.

mindful of spinal cord perfusion and avoid periods of relative hypotension while maintaining these low pressures.

Endovascular Procedures Many of the complications are directly related to manipulation of the delivery system within the iliac arteries and aorta.[146] Patients with small, calcified, tortuous iliofemoral arteries are at particularly high risk for life-threatening iliac artery rupture. Although relatively uncommon, acute iatrogenic retrograde dissection into the aortic arch and ascending aorta is a life-threatening complication that necessitates emergency repair of the ascending aorta and aortic arch via sternotomy and cardiopulmonary bypass.[109] The most important risk factors for this complication include incautious wire and catheter manipulation, aggressive proximal ballooning (especially in cases of acute descending thoracic aortic dissection), and hybrid arch repair in which the native ascending aorta is dilated (more than 4 cm). Retrograde proximal dissection converts a localized descending thoracic aortic aneurysm into an acute problem involving the entire thoracic aorta. Of note, retrograde aortic dissection may also occur several months after initial repair.[147]

Another significant complication of descending thoracic aortic stent grafting is endoleak. An endoleak occurs when there is a persistent flow of blood (visible on radiologic imaging) into the aneurysm sac, and it may occur during the initial procedure or develop over time. Although endoleaks are a relatively common complication,[148,149] they are not benign because they lead to continual pressurization of the sac, which can cause expansion or even rupture. These complications are categorized (Table 22-4) according to the site of the leak.[127] Although all endoleaks may progress such that they can be considered life-threatening, type I and type III endoleaks generally necessitate early and aggressive intervention. Recently published reporting guidelines aid standardized reporting.[127]

Other complications include stent graft misdeployment, device migration, endograft kinking or infolding, and stent graft infection, including fistula. Although not all complications related to stent grafts are fatal, endovascular repairs should be performed by expert teams qualified to address the variety of problems that may arise; some patients may need to have these devices removed and replaced with polyester grafts.[67,68,150,151] Complications of endovascular repair are relatively common, so regularly scheduled radiologic imaging surveillance is of the utmost importance.

Table 22-4

Classification of and common treatment strategies for endoleak

Type I
- Incomplete seal between stent graft and aorta at the proximal landing site (Type Ia), the distal landing site (Type Ib), or branch module, fenestration, or plug (Type Ic)
- Early reintervention to improve seal or conversion to open surgery

Type II
- Retrograde perfusion of sac from excluded collateral arteries
- Surveillance; as-needed occlusion with percutaneous or other interventions

Type III
- Incomplete seal between overlapping stent graft or module (Type IIIa), or tear in graft fabric (Type IIIb)
- Early reintervention to cover or conversion to open surgery

Type IV
- Perfusion of sac due to porosity of material
- Surveillance; as-needed reintervention to reline stent graft

Type V
- Expansion of sac with no identifiable source
- Surveillance; as-needed reintervention to reline stent graft

AORTIC DISSECTION

Pathology and Classification

Aortic dissection, the most common catastrophic event involving the aorta, is a progressive separation of the aortic wall layers that usually occurs after a tear forms in the intima and inner media. As the separation of the layers of the media propagates, two channels are typically formed (Fig. 22-18): the original lumen, which remains lined by the intima and which is called the *true lumen*, and the newly formed channel within the layers of the media, which is called the *false lumen.* The dissecting membrane separates the true and false lumens. Additional tears in the dissecting membrane that allow communication between the two channels are called *reentry sites.* Although the separation of layers primarily progresses distally along the length of the aorta, it can also proceed in a proximal direction; this process often is referred to as *proximal extension* or *retrograde dissection.*

The extensive disruption of the aortic wall has severe anatomic consequences (Fig. 22-19). First, the outer wall of the false lumen is extremely thin, inflamed, and fragile, which makes it prone to expansion or rupture in the face of ongoing hemodynamic stress. Second, the expanding false lumen can compress the true lumen and cause *malperfusion syndrome* by interfering with blood flow in the aorta or any of its branch vessels, including the coronary, carotid, intercostal, visceral, renal, and iliac arteries. Finally, when the separation of layers occurs within the aortic root, the aortic valve commissures can become unhinged, which results in acute valvular regurgitation. The clinical consequences of each of these sequelae are addressed in detail in the section on clinical manifestations.

Dissection vs. Aneurysm. The relationship between dissection and aneurysmal disease requires clarification. Dissection and aneurysm are separate entities, although they often coexist and are mutual risk factors. In some cases, dissection occurs in patients without aneurysms, and the subsequent progressive dilatation of the weakened outer aortic wall ultimately results in an aneurysm. On the other hand, in patients with degenerative aneurysms, the ongoing deterioration of the aortic wall can lead to a superimposed dissection. The overused term *dissecting aneurysm* should be reserved for this specific situation.

Classification. For management purposes, aortic dissections are classified according to their location and chronicity. Improvements in imaging have increasingly revealed variants of aortic dissection that probably represent different forms along the spectrum of this condition.

Location To guide treatment, dissections are categorized according to their anatomic location and extent. The two traditional classification schemes that remain in common use are the DeBakey and the Stanford classification systems (Fig. 22-20).[152,153] In their current forms, both of these schemes describe the segments of aorta that are involved in the dissection, rather than the site of the initial intimal tear. The main drawback of the Stanford classification system is that it does not distinguish between patients with isolated ascending aortic dissection and patients with dissection involving the entire aorta. Both types of patients would be classified as having type A dissections, despite the fact that their treatment, follow-up, and prognosis are substantially different.

Additional classification schemas include that by Borst and associates,[154] in which the ascending and descending aorta are considered independently; the recent modification of the DeBakey classification by Tsagakis et al,[155] which extends type II dissection into the aortic arch; and the Penn modification of the Stanford classification,[156,157] which expands the classification to include the presence of tissue and global malperfusion. These modifications may help to better streamline the primary surgical intervention.

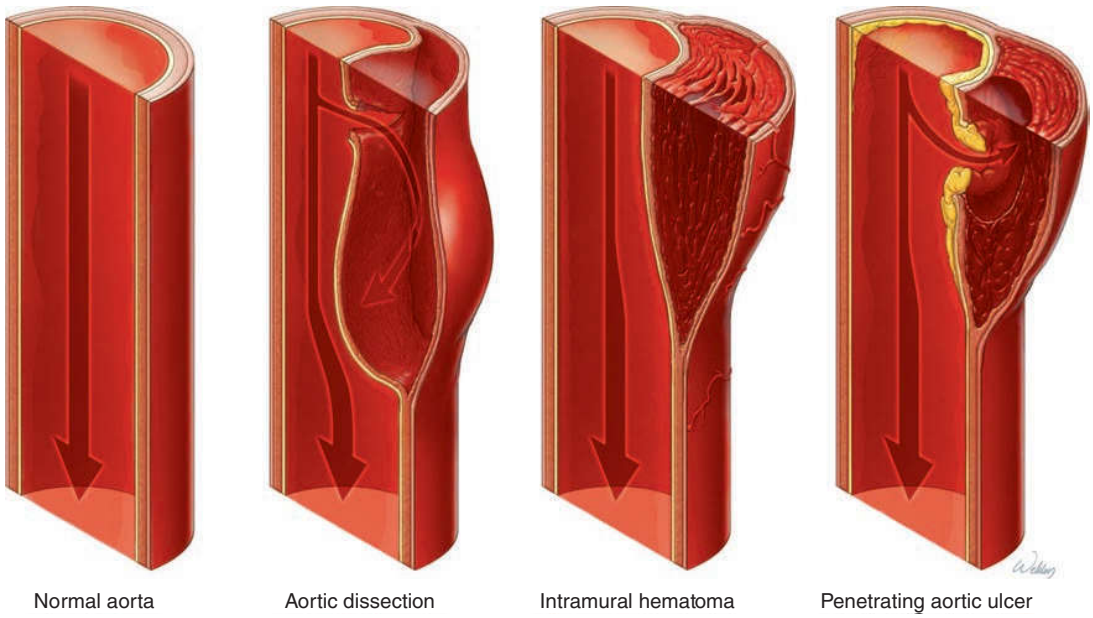


Figure 22-18. Illustration of longitudinal sections of the aortic wall and lumen. Blood flows freely downstream in normal aortic tissue. In classic aortic dissection, blood entering the media through a tear creates a false channel in the wall. Intramural hematomas arise when hemorrhage from the vasa vasorum causes blood to collect within the media; the intima is intact. Penetrating aortic ulcers are deep atherosclerotic lesions that burrow into the aortic wall and allow blood to enter the media. In each of these conditions, the outer aortic wall is severely weakened and prone to rupture.

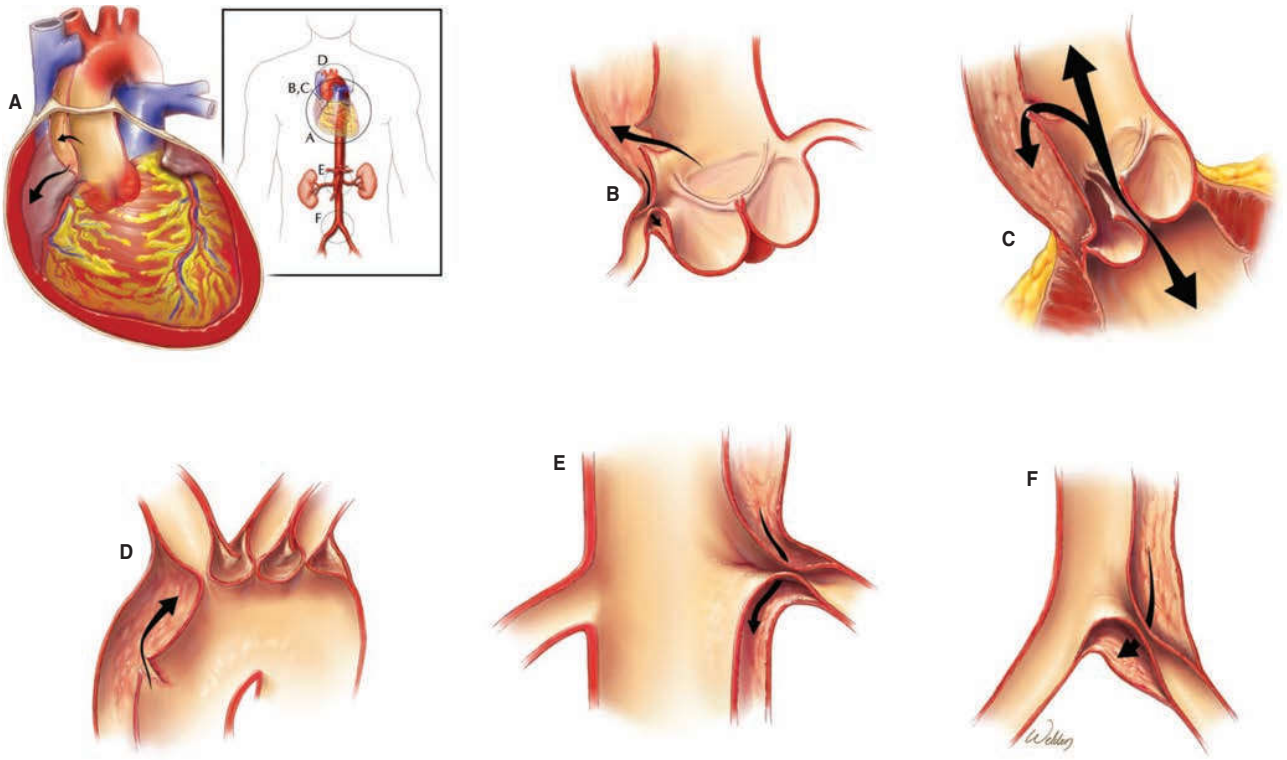


Figure 22-19. Illustration of the potential anatomic consequences of aortic dissection, with a mapped diagram of affected regions (*inset*). **A.** Ascending aortic rupture and cardiac tamponade. **B.** Disruption of coronary blood flow. **C.** Injury to the aortic valve causing regurgitation. **D, E,** and **F.** Compromised blood flow to branch vessels, causing ischemic complications. *(Adapted with permission from Creager MA, Dzau VS, Loscalzo J:* Vascular Medicine*, 7th ed. Philadelphia, PA: Elsevier/Saunders; 2006.)*

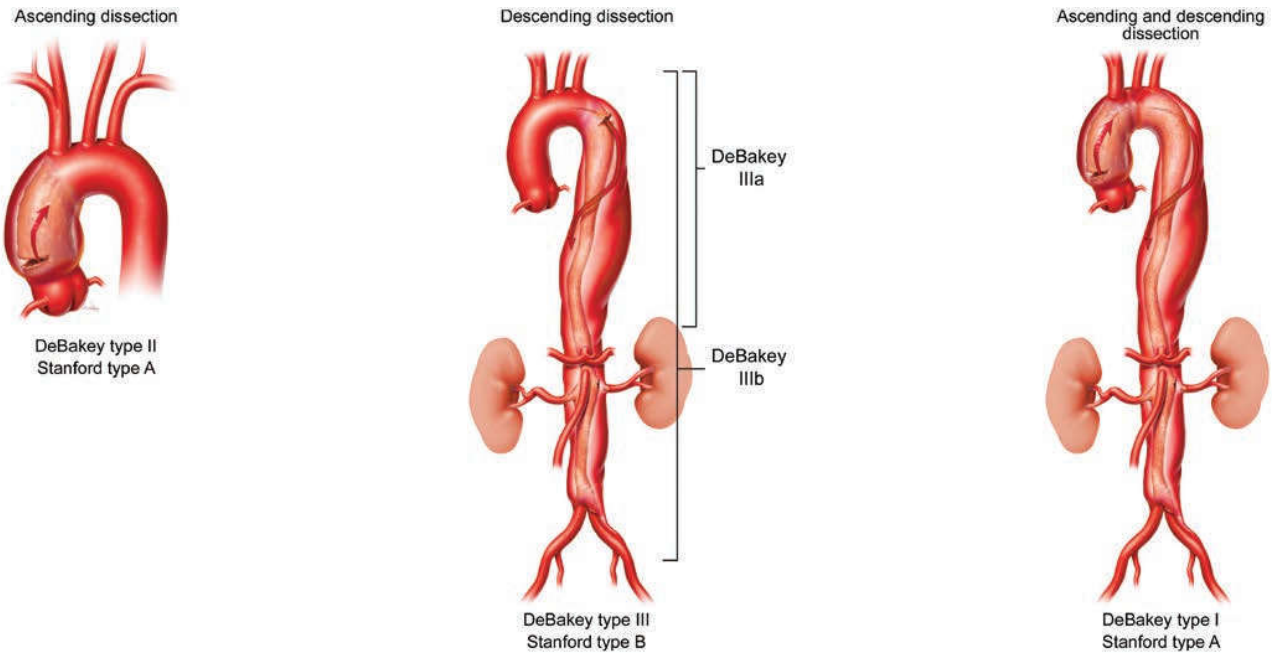


Figure 22-20. Illustration of the classification schemes for aortic dissection based on which portions of the aorta are involved. Dissection can be confined to the ascending aorta (*left*) or the descending aorta (*middle*), or it can involve the entire aorta (*right*). *(Used with permission from Baylor College of Medicine.)*

Regardless of which system is used, patients with isolated ascending aortic dissection usually undergo emergent operation, as do patients with dissection involving both the ascending and descending thoracic segments. Patients with isolated descending thoracic and abdominal aortic dissection are typically treated medically, unless complications requiring surgery develop. Understanding the precise extent of dissection has become increasingly important as some aortic centers consider augmenting traditional ascending aortic repairs with endovascular techniques to treat dissected distal aortic segments.[158]

Chronicity Aortic dissection also is categorized according to the time elapsed since the initial tear. Dissection is considered acute within the first 14 days after the initial tear; after 14 days, the dissection is considered chronic. Although arbitrary, the distinction between acute and chronic dissections has important implications, not only for decision making about perioperative management strategies and operative techniques, but also for evaluating surgical results. Figure 22-21 provides an algorithm for the management of acute aortic dissection. In light of the importance of acuity, Borst and associates[154] have proposed a third phase—termed *subacute*—to describe the transition between the acute and chronic phases. The subacute period encompasses days 15 through 60 after the initial tear. Although this is past the traditional 14-day acute phase, patients with subacute dissection continue to have extremely fragile aortic

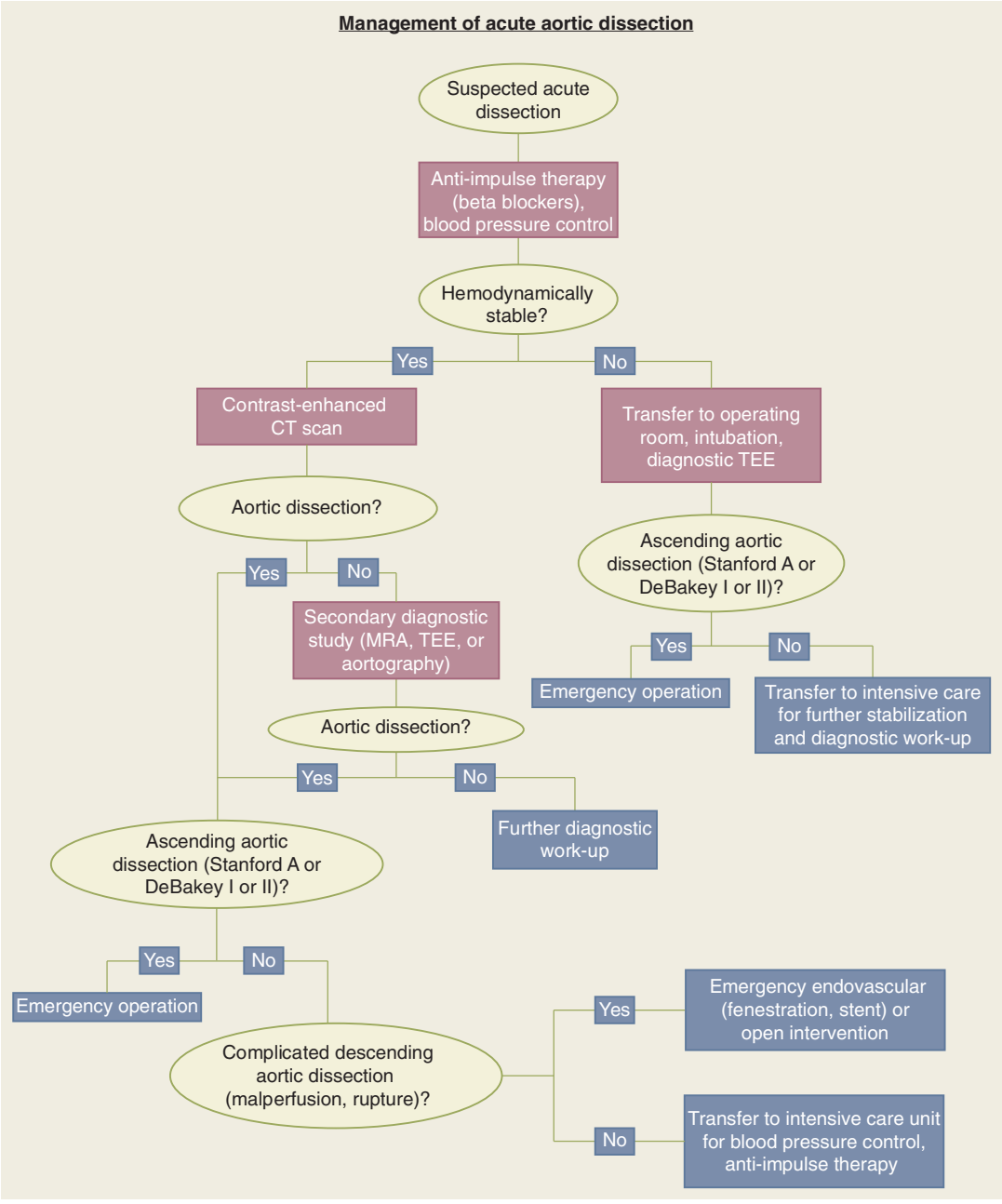


Figure 22-21. Algorithm used to facilitate decisions regarding treatment of acute aortic dissection. CT = computed tomography; MRA = magnetic resonance angiography; TEE = transesophageal echocardiography.

tissue, which may complicate operative treatment and increase the risks associated with surgery. Recently, the International Registry of Acute Aortic Dissections (IRAD) investigators proposed a new classification system for characterizing the phases of aortic dissection: The period within 24 hours from the onset of symptoms was defined as hyperacute, the period between 2 and 7 days was defined as acute, the period between 8 and 30 days was defined as subacute, and the period beyond 30 days was defined as chronic.[159]

Variants As noted earlier, advancements in noninvasive imaging of the aorta have revealed variants of aortic dissection (see Fig. 22-18). The recently introduced term *acute aortic syndrome* encompasses classic aortic dissection and its variants. Other aortic syndromes, which were once thought to be rare, include *intramural hematoma (IMH)* and *penetrating aortic ulcer (PAU).* Although the issue is somewhat controversial, the current consensus is that, in most cases, these variants of dissection should be treated identically to classic dissection.

An IMH is a collection of blood within the aortic wall, without an intimal tear, that is believed to be due to rupture of the vasa vasorum within the media. The accumulation of blood can result in a secondary intimal tear that ultimately leads to a dissection.[160] Because IMH and aortic dissection represent a continuum, it is possible that IMH is seen less frequently than aortic dissection because IMH rapidly progresses to true dissection. The prevalence of IMH among patients with acute aortic syndromes is approximately 6%, and 16% progress to full dissection.[161] An IMH can be classified according to its location (i.e., ascending or descending) and should be treated analogously to classic dissection.[162]

A PAU is essentially a disrupted atherosclerotic plaque that projects into the aortic wall and is associated with surrounding hematoma. Eventually, the ulcer can penetrate the aortic wall, which leads to dissection or rupture. The rate of disease progression is higher than that of IMH alone.[163]

Causes and Clinical History

Aortic dissection is a lethal condition with a reported incidence of 3.5 per 100,000 in the United States.[164] Without appropriate modern medical or surgical treatment, most patients (approximately 90%) die within 3 months of dissection, mostly from rupture.[165,166]

Although several risk factors for aortic dissection have been identified, the specific causes remain unknown. Ultimately, any condition that weakens the aortic wall increases the risk of aortic dissection. Common general cardiovascular risk factors, such as smoking, hypertension, atherosclerosis, and hypercholesterolemia, are associated with aortic dissection. Patients with heritable forms of aortopathy, aortitis, bicuspid aortic valve, or preexisting medial degenerative disease are at risk for dissection, especially if they already have a thoracic aortic aneurysm.[24] Aortic injury during cardiac catheterization, surgery, or endovascular aortic repair is a common cause of iatrogenic dissection. Other conditions that are associated with aortic dissection include cocaine and amphetamine abuse,[167] as well as severe emotional stress or extreme physical exertion such as during weightlifting.[168] Advances in the understanding of the molecular mechanisms behind abdominal aortic aneurysms have prompted similar investigations of thoracic aortic dissection.[169-171]

Clinical Manifestations

The onset of dissection often is associated with severe chest or back pain, classically described as "tearing," that migrates distally as the dissection progresses along the length of the aorta. The location of the pain often indicates which aortic segments are involved. Pain in the anterior chest suggests involvement of the ascending aorta, whereas pain in the back and abdomen generally indicates involvement of the descending and thoracoabdominal aorta. Additional clinical sequelae of acute aortic dissection vary substantially and are best considered in terms of the dissection's potential anatomic manifestations at each level of the aorta (see Fig. 22-19 and Table 22-5). Thus, potential complications of dissection of the aorta (and involved secondary arteries) may include cardiac ischemia (coronary artery) or tamponade, stroke (brachiocephalic arteries), paraplegia or paraparesis (intercostal arteries), mesenteric ischemia (superior mesenteric artery), kidney failure (renal arteries), and limb ischemia or loss of motor function (brachial or femoral arteries).

Ascending aortic dissection can directly injure the aortic valve, causing regurgitation. The severity of the regurgitation varies with the degree of commissural disruption, which ranges from partial separation of only one commissure, producing mild

Table 22-5

Anatomic complications of aortic dissection and their associated symptoms and signs

ANATOMIC MANIFESTATION	SYMPTOMS AND SIGNS
Aortic valve insufficiency	Dyspnea Murmur Pulmonary rales Shock
Coronary malperfusion	Chest pain with characteristics of angina Nausea/vomiting Shock Ischemic changes on electrocardiogram Elevated cardiac enzymes
Pericardial tamponade	Dyspnea Jugular venous distension Pulsus paradoxus Muffled cardiac tones Shock Low-voltage electrocardiogram
Subclavian or iliofemoral artery malperfusion	Cold, painful extremity Extremity sensory and motor deficits Peripheral pulse deficit
Carotid artery malperfusion	Syncope Focal neurologic deficit (transient or persistent) Carotid pulse deficit Coma
Spinal malperfusion	Paraplegia Incontinence
Mesenteric malperfusion	Nausea/vomiting Abdominal pain
Renal malperfusion	Oliguria or anuria Hematuria

valvular regurgitation, to full separation of all three commissures and complete prolapse of the valve into the left ventricle, producing severe acute heart failure. Patients with acute aortic valve regurgitation may report rapidly worsening dyspnea.

Ascending dissections also can extend into the coronary arteries or shear the coronary ostia off of the true lumen, causing acute coronary occlusion; when this occurs, it most often involves the right coronary artery. The sudden disruption of coronary blood flow can cause a myocardial infarction. This presentation of acute myocardial ischemia can mask the presence of aortic dissection, which results in delayed diagnosis and treatment.[172]

The thin and inflamed outer wall of a dissected ascending aorta often produces a serosanguineous pericardial effusion that can accumulate and cause tamponade. Suggestive signs include jugular venous distention, muffled heart tones, pulsus paradoxus, and low-voltage electrocardiogram (ECG) tracings. Free rupture into the pericardial space produces rapid tamponade and is generally fatal.

As the dissection progresses, any branch vessel from the aorta can become involved, which results in compromised blood flow and ischemic complications (i.e., *malperfusion*). Therefore, depending on which arteries are involved, the dissection can produce acute stroke, paraplegia, hepatic failure, bowel infarction, renal failure, or a threatened ischemic limb.

Diagnostic Evaluation

Because of the variations in severity and the wide variety of potential clinical manifestations, the diagnosis of acute aortic dissection can be challenging.[173-175] Only 3 out of every 100,000 patients who present to an emergency department with acute chest, back, or abdominal pain are eventually diagnosed with aortic dissection. Not surprisingly, diagnostic delays are common; delays beyond 24 hours after hospitalization occur in up to 39% of cases. Unfortunately, delays in diagnosis lead to delays in treatment, which can have disastrous consequences. The European Society of Cardiology Task Force on Aortic Dissection stated, "The main challenge in managing acute aortic dissection is to suspect and thus diagnose the disease as early as possible."[173] A recent study by the IRAD investigators examined the reasons for delayed diagnosis and found that diagnosis lagged in women, as well as in patients with atypical symptoms, such as fever or mild pain (rather than severe pain).[172] A high index of suspicion is critical, particularly in younger, atypical patients, who may have heritable disorders or other, less common risk factors.

Most patients with acute aortic dissection (80% to 90%) experience severe pain in the chest, back, or abdomen.[173-175] The pain usually occurs suddenly, has a sharp or tearing quality, and often migrates distally as the dissection progresses along the aorta. For classification purposes (acute vs. subacute vs. chronic), the onset of pain is generally considered to represent the beginning of the dissection process. Most of the other common symptoms either are nonspecific or are caused by the secondary manifestations of dissection.

A discrepancy between the extremities in pulse, blood pressure, or both is the classic physical finding in patients with aortic dissection. It often occurs because of changes in flow in the true and false lumens, and it does not necessarily indicate extension into an extremity branch vessel. Involvement of the aortic arch often creates differences between the right and left arms, whereas descending aortic dissection often causes differences between the upper and lower extremities. Like symptoms, most of the physical signs after dissection are related to the secondary manifestations and therefore vary considerably (see Table 22-5). For example, signs of stroke or a threatened ischemic limb may dominate the physical findings in patients with carotid or iliac malperfusion, respectively.

Unfortunately, laboratory studies are of little help in diagnosing acute aortic dissection. There has been continued interest in using D-dimer level to aid in making this diagnosis.[176] Several reports indicate that D-dimer is an extremely sensitive indicator of acute aortic dissection; elevated levels are found in approximately 97% of affected patients.[177] Tests that are commonly used to detect acute coronary events—including ECG and tests for serum markers of myocardial injury—deserve special consideration and need to be interpreted carefully. Normal ECGs and serum marker levels in patients with acute chest pain should raise suspicion about the possibility of aortic dissection. It is important to remember that ECG changes and elevated serum marker levels associated with myocardial infarction do not exclude the diagnosis of aortic dissection because dissection can cause coronary malperfusion. Of note, abnormal ECGs have recently been shown to delay the diagnosis of aortic dissection, and the possibility of aortic dissection should not be prematurely ruled out.[172,178] Similarly, although CXRs may show a widened mediastinum or abnormal aortic contour, up to 16% of patients with dissection have a normal-appearing CXR.[174] The value of the CXR for detecting aortic dissection is limited, with a sensitivity of 67% and a specificity of 86%.[179]

Once the diagnosis of dissection is considered, the thoracic aorta should be imaged with CT, MRA, or echocardiography. The accuracy of these noninvasive imaging tests has all but eliminated the need for diagnostic aortography in most patients with suspected aortic dissection. Currently, the diagnosis of aortic dissection is usually established with contrast-enhanced CT, which has a sensitivity of 98% and a specificity of 87%, and, most importantly, acquires images swiftly.[180] The classic diagnostic feature is a double-lumen aorta (Fig. 22-22). In addition, CT scans provide essential information about the segments of the aorta involved; the acuity of the dissection; aortic dilatation, including the presence of preexisting degenerative aneurysms; and the development of threatening sequelae, including pericardial effusion, early aortic rupture, and branch-vessel compromise. Although MRA also provides excellent imaging (with both a sensitivity and specificity of 98%), the MR suite is not well suited for critically ill patients. In patients who cannot undergo contrast-enhanced CT or MRA, transthoracic echocardiography can be used to establish the diagnosis.

Transesophageal echocardiography (TEE) is excellent for detecting dissection, aneurysm, and IMH in the ascending aorta. In appropriate hands, TEE has a demonstrated sensitivity and specificity as high as 98% and 95%, respectively.[181] Furthermore, TEE offers important information about ventricular function and aortic valve competency. Finally, TEE is the diagnostic modality of choice for hemodynamically unstable patients in whom the diagnosis of ascending dissection is suspected; ideally, these patients should be taken to the operating room, where the TEE can be performed and, if the TEE is confirmatory, surgery can be started immediately.

In selected patients with ascending aortic dissection (i.e., those who have evidence of preexisting coronary artery disease), coronary angiography can be considered before surgery. Specific relative indications in these patients include a history

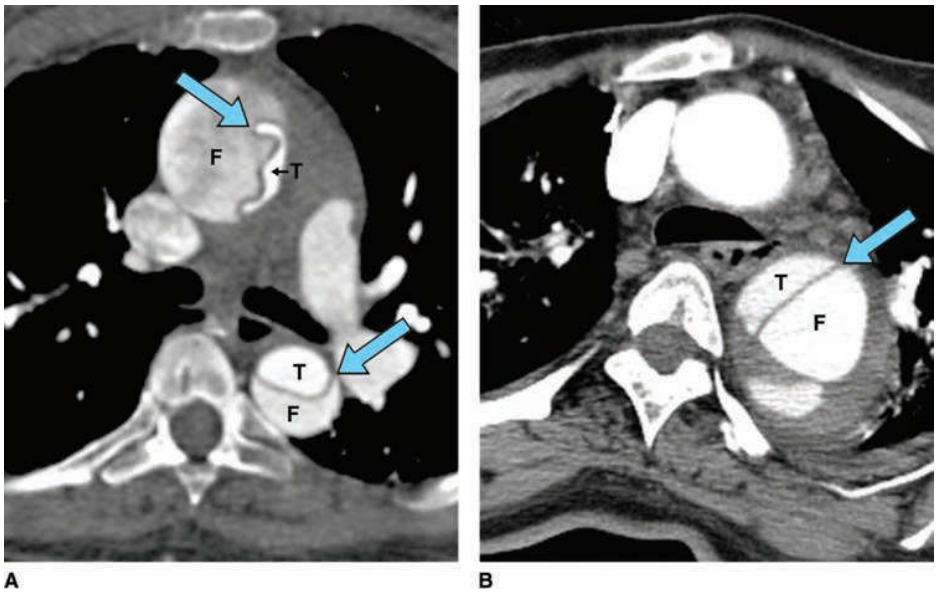


Figure 22-22. Computed tomographic scans showing that the aorta has been separated into two channels—the true (T) and false (F) lumens—in two patients with different phases of aortic dissection. **A.** An acute DeBakey type I aortic dissection. The dissecting membrane appears wavy (*arrows*) in the early phase of dissection. Here, the true lumen of the proximal aorta can be seen to be extensively compressed. This may lead to malperfusion of the heart. **B.** A chronic DeBakey type III aortic dissection. In the chronic phase, the membrane appears straighter and less mobile (*arrow*) because it has stabilized over time. *(Used with permission of Baylor College of Medicine.)*

of angina or myocardial infarction, a recent myocardial perfusion study with abnormal results, previous coronary artery bypass or angioplasty, and acute ischemic changes on ECG. Contraindications include hemodynamic instability, aortic rupture, and pericardial effusion.[182] In our practice, patients with acute aortic dissections rarely undergo coronary angiography. However, all patients presenting for elective repair of chronic ascending dissections have diagnostic coronary angiograms taken.

Of note, when malperfusion of the renal, visceral, or lower extremity arteries develops, the patient is usually treated in an angiography suite or hybrid operating room.[158] Although the dissection usually is diagnosed on CT scan, these patients also undergo aortography, during which the mechanism of the malperfusion is ascertained and, if possible, corrected. Hence, catheter-based aortography may be obsolete as a diagnostic test for dissection, but it remains beneficial for patients with malperfusion.

Treatment

Initial Assessment and Management. Regardless of the location of the dissection, the initial treatment is the same for all patients with suspected or confirmed acute aortic dissection (see Fig. 22-21). Furthermore, because of the potential for rupture before the diagnosis is confirmed, aggressive pharmacologic management is started once there is clinical suspicion of dissection, and this treatment is continued during the diagnostic evaluation. The goals of pharmacologic treatment are to stabilize the dissection and prevent rupture.

Patients are monitored closely in an intensive care unit. Indwelling radial arterial catheters are used to monitor blood pressure and optimize titration of antihypertensive agents. Blood pressures in a malperfused limb can underrepresent the central aortic pressure; therefore, blood pressure is measured in the arm with the better pulse. Central venous catheters assure reliable IV access for delivering vasoactive medications. Pulmonary artery catheters are reserved for patients with severe cardiopulmonary dysfunction.

In addition to confirming the diagnosis of dissection and defining its acuity and extent, the initial evaluation focuses on determining whether any of several life-threatening complications are present. Particular attention is paid to changes in neurologic status, peripheral pulses, and urine output. Serial laboratory studies—including arterial blood gas concentrations, complete blood cell count, prothrombin and partial thromboplastin times, and serum levels of electrolytes, creatinine, blood urea nitrogen, and liver enzymes—are useful for detecting organ ischemia and optimizing management.

The initial management strategy, commonly described as *anti-impulse therapy* or *blood pressure control,* focuses on reducing aortic wall stress, the force of left ventricular ejection, chronotropy, and the rate of change in blood pressure (dP/dT). Reductions in dP/dT are achieved by lowering both cardiac contractility and blood pressure. The drugs initially used to accomplish these goals include IV β-adrenergic blockers, direct vasodilators, calcium channel blockers, and angiotensin-converting enzyme inhibitors. These agents are used to achieve a heart rate between 60 and 80 bpm, a systolic blood pressure between 100 and 110 mmHg, and a mean arterial blood pressure between 60 and 75 mmHg. These hemodynamic targets are maintained as long as urine output remains adequate and neurologic function is not impaired. Achieving adequate pain control with IV opiates, such as morphine and fentanyl, is important for maintaining acceptable blood pressure control.

β-Antagonists are administered to all patients with acute aortic dissections unless there are strong contraindications, such as severe heart failure, bradyarrhythmia, high-grade atrioventricular conduction block, or bronchospastic disease. Esmolol

can be useful in patients with bronchospastic disease because it is a cardioselective, ultra-fast-acting agent with a short half-life. Labetalol, which causes both nonselective β-blockade and postsynaptic α_1-blockade, reduces systemic vascular resistance without impairing cardiac output. Doses of β-antagonists are titrated to achieve a heart rate of 60 to 80 bpm. In patients who cannot receive β-antagonists, calcium channel blockers such as diltiazem are an effective alternative. Nitroprusside, a direct vasodilator, can be administered once β-blockade is adequate. When used alone, however, nitroprusside can cause reflex increases in heart rate and contractility, elevated dP/dT, and progression of aortic dissection. Enalapril and other angiotensin-converting enzyme inhibitors are useful in patients with renal malperfusion. These drugs inhibit renin release, which may improve renal blood flow.

Treatment of Ascending Aortic Dissection

Acute Dissection Because of the risk of aortic rupture, acute ascending aortic dissection is usually considered an absolute indication for emergency surgical repair. However, specific patient groups may benefit from nonoperative management or delayed operation.[183] Delayed repair may be considered for patients who (a) present with severe acute stroke or mesenteric ischemia, (b) are elderly and have substantial comorbidity, (c) are in stable condition and may benefit from transfer to specialized centers, or (d) have undergone a cardiac operation in the remote past. Regarding the last group, it is important that the previous operation not be too recent; dissections that occur during the first 3 weeks after cardiac surgery pose a high risk of rupture and tamponade, and such dissections warrant early operation.[184]

In the absence of the aforementioned circumstances, most patients with acute ascending aortic dissection undergo emergent graft replacement of the ascending aorta. Operative repair is similar to that for aneurysm of the transverse aortic arch (previously described) because hypothermic circulatory arrest is commonly used regardless of the extent of repair. Immediately before the operation begins, intraoperative TEE is commonly performed to further assess baseline myocardial and valvular function and, if necessary, to confirm the diagnosis. The operation is performed via a median sternotomy with cardiopulmonary bypass and hypothermic circulatory arrest (Fig. 22-23). In preparation for circulatory arrest, cannulas are placed in the right axillary artery (to provide arterial inflow) and in the right atrium (to provide venous drainage).[94] The innominate artery is sometimes used for arterial inflow if it is not dissected.[185] After an appropriate level of cooling has been achieved (approximately 24°C), cardiopulmonary bypass is stopped, and the ascending aorta is opened. The innominate artery is then occluded with a clamp or snare, and flow from the axillary artery cannula is used to provide ACP.[186] Currently as a default, we use bilateral ACP with a separate perfusion catheter in the left common carotid artery to ensure perfusion of the left side of the brain. This strategy of performing the distal anastomosis during a brief period of circulatory arrest, often termed *open distal anastomosis*, obviates the need to place a clamp across the fragile aorta, avoiding further aortic damage. Also, it allows the surgeon to carefully inspect the aortic arch for intimal tears. Traditionally, the entire arch is replaced only if a primary intimal tear is located in the arch or if the arch is aneurysmal; most commonly, repair is limited to replacement of the entire ascending aorta or to a beveled "hemiarch" repair.[187] Conservative repair has been shown to increase the likelihood of early survival.[188] The distal aortic cuff is prepared by tacking the inner and outer walls together and occasionally using a small amount of surgical adhesive to obliterate the false lumen and strengthen the tissue. A polyester tube graft is sutured to the distal aortic cuff. The anastomosis between the graft and the aorta is fashioned so that blood flow will be directed into the true lumen; this often alleviates distal malperfusion problems that were present preoperatively. After the distal anastomosis has been completed and adequately reinforced, the graft is deaired and clamped, full cardiopulmonary bypass is resumed, rewarming is initiated, and the proximal portion of the repair is started. In the absence of conditions that generally necessitate aortic root replacement (i.e., annuloaortic ectasia or heritable disorders, particularly Marfan and Loeys-Dietz syndromes), aortic valve regurgitation can be corrected by resuspending the commissures onto the outer aortic wall.[189] The proximal aortic cuff is prepared with tacking sutures and occasionally a small amount of surgical adhesive before the proximal aortic anastomosis is performed.

In the majority of patients who undergo surgical repair of acute ascending dissection, the dissection persists distal to the site of the operative repair; the residually dissected aorta, which generally includes at least a portion of the transverse aortic arch as well as a large portion of the distal aorta, is susceptible to dilatation over time. Extensive dilatation of the arch or distal aorta develops in 25% to 40% of survivors[190,191] and often necessitates further aortic repair. Additionally, long-term survival after acute proximal aortic dissection is generally poor, and rupture of the dilated distal aorta is a common cause of late death in these patients.[188,190-192]

The challenges that survivors of acute proximal aortic dissection commonly face over time have led to the development of alternate acute dissection strategies such as total arch replacement[193] and hybrid arch strategies to extend proximal aortic repair into the distal aorta. The goal of hybrid arch approaches in acute dissection is to thrombose the residual false lumen by compressing it with the radial force that is exerted by a stent graft placed in the true lumen, thereby facilitating remodeling and preventing late aneurysm formation.[194,195] However, in such repairs, the compressed false lumen may continue to be perfused in a retrograde fashion.

In Europe, Japan, and elsewhere, one-piece hybrid prostheses are now available that incorporate a polyester graft for the proximal repair and a stent graft component for the descending aorta. The device enables single-stage "frozen elephant trunk" repair of the ascending aorta, entire aortic arch, and proximal descending thoracic aorta.[196] In the United States, such devices are not currently available, so this repair is commonly done by concomitantly deploying a commercially available stent graft in an antegrade fashion after fully replacing the ascending aorta and aortic arch. In some variations of this off-label approach, the stent graft is directly sutured to the distal aspect of the proximal open repair, whereas in others, there may be a gap of native tissue between the open and endovascular repair. Although this technique appears to be extensively used outside the United States, and with early and mid-term success,[194,197-199] only a few U.S. reports describe its use.[200-203] Emerging reports describe an enhanced risk of spinal cord ischemia, a risk that is not usually associated with open arch repair. This is probably due to the extensive coverage of the intercostal vessels by the stent graft. Uncertainties in the frozen elephant trunk procedure need to be addressed before it becomes a standard

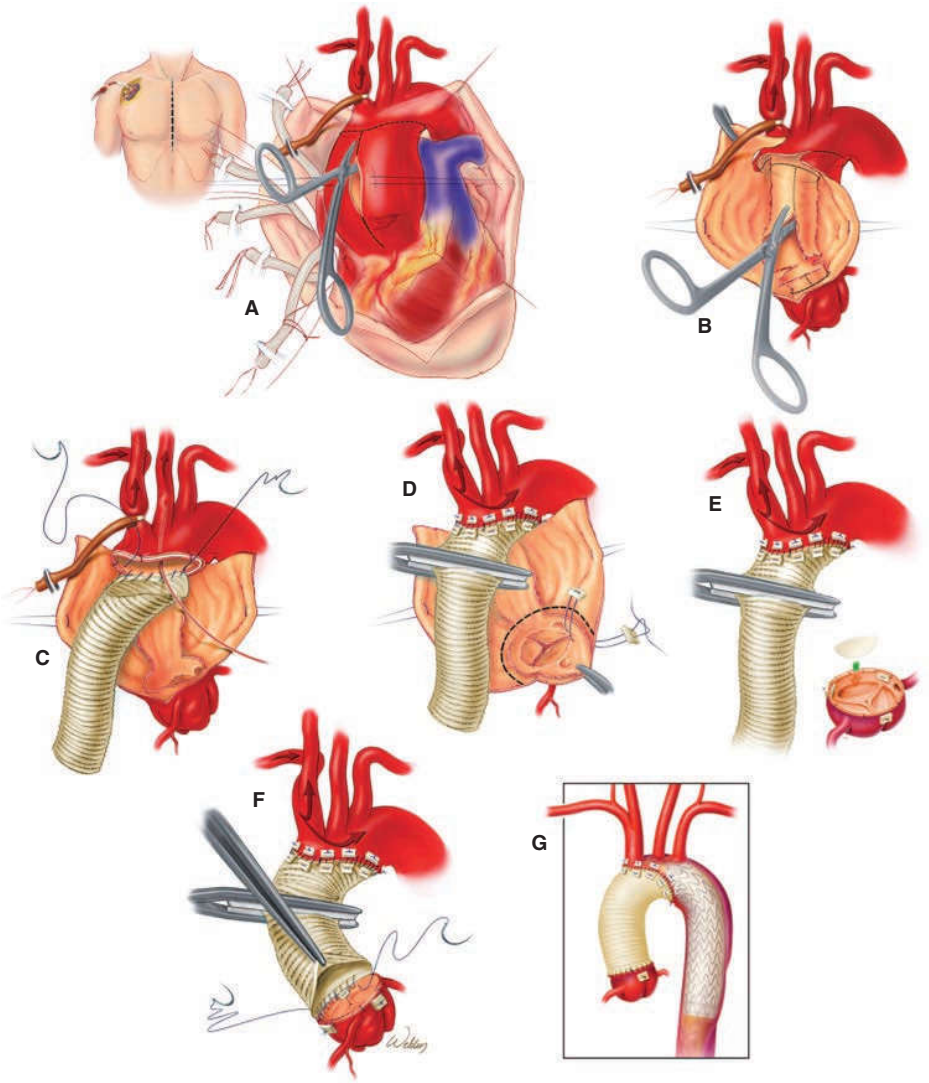


Figure 22-23. Illustration of proximal aortic repair for acute ascending aortic dissection. **A.** This repair requires a median sternotomy and cardiopulmonary bypass. The ascending aorta is opened during hypothermic circulatory arrest, while antegrade cerebral perfusion is delivered via an axillary artery graft (shown) or via an innominate artery graft, provided that the innominate artery is not dissected (see Fig. 22-10). **B.** The dissecting membrane is removed to expose the true lumen. **C.** An open distal anastomosis prevents clamp injury of the friable arch tissue and allows inspection of the arch lumen. A balloon perfusion catheter in the left common carotid artery ensures bilateral antegrade cerebral perfusion. If the origin of the dissection (i.e., intimal tear or disruption) does not extensively involve the greater curvature of the aortic arch, and if there is no evidence of a preexisting arch aneurysm, a beveled, hemiarch repair is carried out, preserving most of the greater curvature of the arch. The aorta is transected, beginning at the greater curvature immediately proximal to the origin of the innominate artery and extending distally toward the lesser curvature to the level of the left subclavian artery. Consequently, most of the transverse aortic arch, except for the dorsal segment containing the brachiocephalic arteries, is removed. An appropriately sized, sealed (with collagen or gelatin) polyester tube graft is selected, and the beveled distal anastomosis is made with continuous 3-0 or 4-0 monofilament suture; the potential space between the true and false lumen can be obliterated with a small amount of surgical adhesive or by using a strip of Teflon felt. To improve hemostasis, the distal anastomosis can be reinforced by placing interrupted mattress sutures with felt pledgets. **D.** After cardiopulmonary bypass is resumed and a cross-clamp is applied to the hemiarch replacement graft, the aortic valve is assessed. Disrupted commissures are resuspended with pledgeted mattress sutures to restore valvular competence. **E.** The aorta is generally transected at the sinotubular junction, and a very small amount of surgical adhesive can be applied between the true and false lumens, or more commonly, the false lumen within the proximal aortic stump is obliterated by inserting a semicircle of felt within the false lumen of the noncoronary sinus. The trimmed edges are brought together by using 6-0 polypropylene sutures. **F.** The proximal anastomosis is carried out at the sinotubular junction, incorporating the distal margin of the commissures. **G.** In patients with residual distal aortic dissection (such as in DeBakey type I aortic dissection), hemiarch repair can be extended with antegrade stent delivery to the descending thoracic aorta. *(Used with permission of Baylor College of Medicine.)*

recommendation for this subset of patients.[204] Another alternative employs separate grafts: a standard polyester graft to replace the ascending aorta and proximal hemiarch, and a stent graft delivered antegrade into the descending thoracic aorta (Fig. 22-23G). Although this procedure differs from a formal "frozen elephant trunk" repair in that it does not replace the entire arch, it is meant to achieve the same goal: promoting remodeling of the dissected descending aortic segment.

Chronic Dissection Occasionally, patients with ascending aortic dissection present for repair in the chronic phase. In most respects, the operation is similar to that for acute dissection repair. One notable difference is that the tissue is stronger in chronic dissection than in acute dissection, which makes suturing safer. In addition, the false lumen is not obliterated at the distal anastomosis; instead, the dissecting membrane is fenestrated into the arch to assure perfusion of both lumens and to prevent postoperative malperfusion complications. Unlike operations for acute dissection, operations for chronic dissection are often aggressive repairs that extend into the arch and root because the tissues are much less fragile.

Treatment of Descending Aortic Dissection

Nonoperative Management Nonoperative, pharmacologic management of acute descending aortic dissection results in lower morbidity and mortality rates than traditional open surgical treatment does.[174] The most common causes of death during nonoperative treatment are aortic rupture and end-organ malperfusion. Therefore, patients are continually reassessed for new complications. Serial CT scans are generally obtained during the index hospitalization—usually on day 2 or 3 and on day 8 or 9 of treatment—and compared with the initial scan to rule out significant aortic expansion.

Once the patient's condition has been stabilized, pharmacologic management is gradually shifted from IV to oral medications. Oral therapy, usually including a β-antagonist, is initiated when systolic pressure is consistently between 100 and 110 mmHg and the neurologic, renal, and cardiovascular systems are stable. Many patients can be discharged after their blood pressure is well controlled with oral agents and after serial CT scans confirm the absence of aortic expansion.

Long-term pharmacologic therapy is important for patients with chronic aortic dissection. β-Blockers remain the drugs of choice.[205] In a 20-year follow-up study, DeBakey and colleagues[206] found that inadequate blood pressure control was associated with late aneurysm formation. Aneurysms developed in only 17% of patients with "good" blood pressure control, compared with 45% of patients with "poor" control.

Aggressive imaging follow-up is recommended for all patients with chronic aortic dissection.[207] Both contrast-enhanced CT and MRA scans provide excellent aortic imaging and facilitate serial comparisons to detect progressive aortic expansion. The first surveillance scan is obtained approximately 6 weeks after the onset of dissection. Subsequent scans are obtained at 3 to 6 months and then at 1 year after onset. If the aorta appears to be stable, imaging is obtained annually thereafter. Scans are obtained more frequently in high-risk patients, such as those with Marfan or Loeys-Dietz syndrome, and in those in whom significant aortic expansion is detected. For patients who have undergone graft repair of descending aortic dissection, annual CT or MRA scans are also obtained to detect false aneurysm formation or dilatation of unrepaired segments of aorta. Early detection of worrisome changes allows timely, elective intervention before rupture or other complications develop; rupture of the distal aorta is relatively common in patients with chronic aortic dissection and often results in death.[192]

Indications for Open Surgery In the acute phase of descending aortic dissection, open surgery has been traditionally reserved for patients who experience complications.[208] Complicated acute distal aortic dissections are those with aortic rupture, increasing periaortic or pleural fluid volume, rapidly expanding aortic diameter, uncontrolled hypertension, and persistent pain despite adequate medical therapy and malperfusion. In general terms, emergency open operations were originally intended to prevent or repair rupture and relieve life-threatening ischemic manifestations. However, open operation is associated with high morbidity in such cases; now that stent graft technology is available, endovascular surgical intervention is recommended for patients with complicated acute distal aortic dissection.

Acute dissection superimposed on a preexisting aneurysm is considered a life-threatening condition and is therefore another indication for operation. Finally, patients who have a history of noncompliance with medical therapy may ultimately benefit more from surgical intervention if they are otherwise reasonable operative candidates.

In the chronic phase, the indications for open surgical intervention for aortic dissection are similar to those for degenerative thoracic aortic aneurysm, although a slightly lower threshold of repair is now recommended. Guidelines for thoracic aortic disease[44] recommend elective operation in otherwise healthy patients when the affected segment has reached a diameter of 5.5 cm, especially in patients with heritable disorders. Rapid aortic enlargement and other factors that increase the likelihood of aortic rupture may also be considered.

Endovascular Treatment

Malperfusion Syndrome Endovascular therapy is routinely used in patients with descending aortic dissection complicated by visceral malperfusion.[209] Abdominal malperfusion syndrome often is fatal; prompt identification of visceral ischemia and expedited treatment to restore hepatic, gastrointestinal, and renal perfusion are imperative for a positive outcome. As described in a later section, several open surgical techniques can be used to reestablish blood flow to compromised organs. However, in acute cases, open surgery is associated with poor outcomes. Therefore, endovascular intervention is the preferred initial approach in such cases. In one endovascular technique known as *endovascular fenestration*, a balloon is used to create a tear in the dissection flap, which allows blood to flow in both the true and false lumens. Although endovascular fenestration was commonly used in the past, its use has declined in recent years as direct aortic and branch-vessel stenting techniques have evolved and gained favor. Placing a stent graft in the true lumen of the aorta can resolve a "dynamic" malperfusion. Occasionally, a small stent must be placed directly in the lumen of a visceral or renal artery because the dissection has propagated into the branch, resulting in "static" malperfusion at the origin.[209]

Iliofemoral malperfusion causing limb-threatening leg ischemia also can be treated via an endovascular approach. Limb malperfusion usually resolves after the endovascular repair of acute descending thoracic aortic dissection. If the malperfusion does not resolve, then a femoral-to-femoral arterial bypass graft is an effective option.

Acute Dissection Although surgery has been traditionally recommended for patients with complicated acute descending aortic dissection, many centers have shifted toward using endovascular stent grafts as the preferred approach in these cases because of the high morbidity associated with the open operation. Evidence suggests that emergent endovascular repair in patients with true lumen collapse and complications such rupture or dynamic malperfusion may be lifesaving in these difficult-to-treat patients. However, these patients remain at risk of further complication or future reintervention. Although endovascular repair in patients with heritable aortic disorders is generally not recommended, this technique can be used as a bridge to later, definitive repair in such life-threatening circumstances.[66]

Controversy exists regarding the use of endovascular stent grafts to treat uncomplicated acute descending dissection; some encouraging data have been published in the last couple of years.[210] The goal of this treatment strategy is to use the stent graft to cover the intimal tear, seal the entry site of the dissection, and eventually cause thrombosis of the false lumen to aid in aortic remodeling and reduce late aortic expansion. Such procedures take place in a hybrid operating room. After the true lumen is accessed through the femoral arteries, an aortogram is taken, and the intimal tear is identified. Note that the diameter of the true lumen is measured on the preoperative contrast-enhanced CT scan. The use of IVUS is encouraged to help access the true lumen, verify navigation of the wire inside the true lumen, and confirm measurements. For these cases, a stent graft is selected with a diameter no more than 10% greater than that of the true lumen. Unlike stents deployed to treat most descending thoracic aortic aneurysms, stents deployed to treat descending thoracic aortic dissections must *not* be ballooned, because ballooning can cause a new intimal tear, retrograde dissection into the ascending aorta, or even aortic rupture. The ideal length of the descending thoracic aorta that should be covered in patients with acute distal descending dissection remains unclear. Close monitoring with serial imaging is necessary after endovascular repair because the false lumen remains at risk for retrograde perfusion or pressurization.

Chronic Dissection Endovascular treatment of chronic descending aortic dissection is supported by the 5-year data of the INSTEAD-XL trial, which showed that endovascular repair combined with optimal medical treatment was associated with slower disease progression and greater aorta-specific survival than optimal medical treatment alone.[211] Importantly, patients in the trial had dissections in the early chronic phase, many within 10 to 12 weeks of onset and all within 1 year of onset. Endovascular repair of chronic dissection is particularly challenging because the relative rigidity of the dissecting membrane—which increases over time during the chronic phase—and the presence of multiple reentry sites make it difficult to exclude the false lumen.

Penetrating Aortic Ulcer Patients with PAUs appear to be good candidates for endovascular intervention. Covering the focal ulceration with a stent graft has been shown to be an effective treatment.[212] In a recent study by Patel and colleagues,[213] endovascular repair of PAU was associated with better early outcomes than open repair. However, when PAU was associated with adjacent hematoma within the aortic wall, rates of subsequent reintervention were increased.

Open Repair

Acute Dissection In patients with acute aortic dissection, open surgical repair of the descending thoracic or thoracoabdominal aorta has been traditionally associated with high morbidity and mortality.[174] Therefore, surgery was generally only performed to prevent fatal rupture or to restore branch-vessel perfusion in patients with complicated dissection.[208] With the evolution of endovascular technology, open repair has fallen out of favor in recent years.

Malperfusion Syndrome In patients with malperfusion, when an endovascular approach is unavailable or unsuccessful, open surgery is necessary. Lower-extremity ischemia can be readily addressed with surgical extra-anatomic revascularization techniques, such as femoral-to-femoral bypass grafting. In patients with abdominal organ ischemia, flow to the compromised bed must be reestablished swiftly. Although they are considered second-line therapies, multiple techniques are available, including graft replacement of the aorta (with flow redirected into the true lumen), open aortic fenestration, and visceral or renal artery bypass.

Chronic Dissection A more aggressive replacement usually is performed during elective aortic repairs in patients with chronic dissection. In many regards, the operative approach used in these patients is identical to that used for descending thoracic and thoracoabdominal aortic aneurysms, as described in the first half of this chapter (Fig. 22-24). One key difference is the need to excise as much dissecting membrane as possible to clearly identify the true and false lumens and to locate all important branch vessels. When the dissection extends into the visceral or renal arteries, the membrane can be fenestrated, or the false lumen can be obliterated with sutures or intraluminal stents. Asymmetric expansion of the false lumen can create wide separation of the renal arteries. This problem is addressed by reattaching the mobilized left renal artery to a separate opening in the graft or by performing a left renal artery bypass with a side graft. Wedges of dissecting membrane also are excised from the aorta adjacent to the proximal and distal anastomoses, which allows blood to flow through both true and false lumens. When placing the proximal clamp is not technically feasible, hypothermic circulatory arrest can be used to facilitate the proximal portion of the repair.

OUTCOMES

Improvements in anesthesia, surgical techniques, and perioperative care have led to substantial improvements in outcome after thoracic aortic aneurysm repair. When performed in specialized centers, these operations are associated with excellent survival rates and acceptable morbidity rates. The interpretation of outcomes data is complicated by site-specific variables, such as the number of years reported and whether data are taken from single-practice centers or from pooled, multicenter, or national registries, and by patient-specific variables, such as type of enrollment, urgency and extent of repair, concomitant procedures performed, and the presence of preexisting risk factors such as advanced age, previous cardiovascular repair, disease of any system or organ, or heritable conditions.

Repair of Proximal Aortic Aneurysms

Risks associated with the open repair of the proximal aorta vary by extent of repair and are greatest for repairs involving total arch replacement.[71,214] All varieties of aortic root replacement have shown acceptable early mortality rates and few complications. Two groups with 20 and 27 years' experience with composite valve graft replacement reported early mortality rates of 5.6% and 1.9%, respectively; the more recent repairs had better outcomes.[215,216] Early mortality rates for stentless porcine

tissue root replacements are also low, ranging from 3.6% to 6.0%.[217-221] Early mortality rates for contemporary valve-sparing approaches to aortic root replacement are quite low (1%–2%) in experienced centers.[77,78,80,222,223] Late survival rates after valve-sparing root procedures range from 97% to 99% at 5 years[80,222,223] and approach 94% at 10 years.[80]

Repairs incorporating the ascending aorta and aortic arch have acceptable outcomes; risk increases with patient-specific factors such as severe atherosclerosis[224] or as larger sections of the aortic arch are incorporated into the repair.[225,226] A revised surgical strategy—such as the use of hypothermic circulatory arrest—is often needed to avoid clamping atherosclerotic sections in the "porcelain" aorta. In Zingone and colleagues' series[224] of 64 patients who underwent replacement of atherosclerotic ascending aorta, hypothermic circulatory arrest was used in 61 patients (95%). Even though these patients had substantial comorbidity and 83% underwent concomitant cardiac repairs, acceptable rates of early mortality (11%) and stroke (6%) were obtained. Other

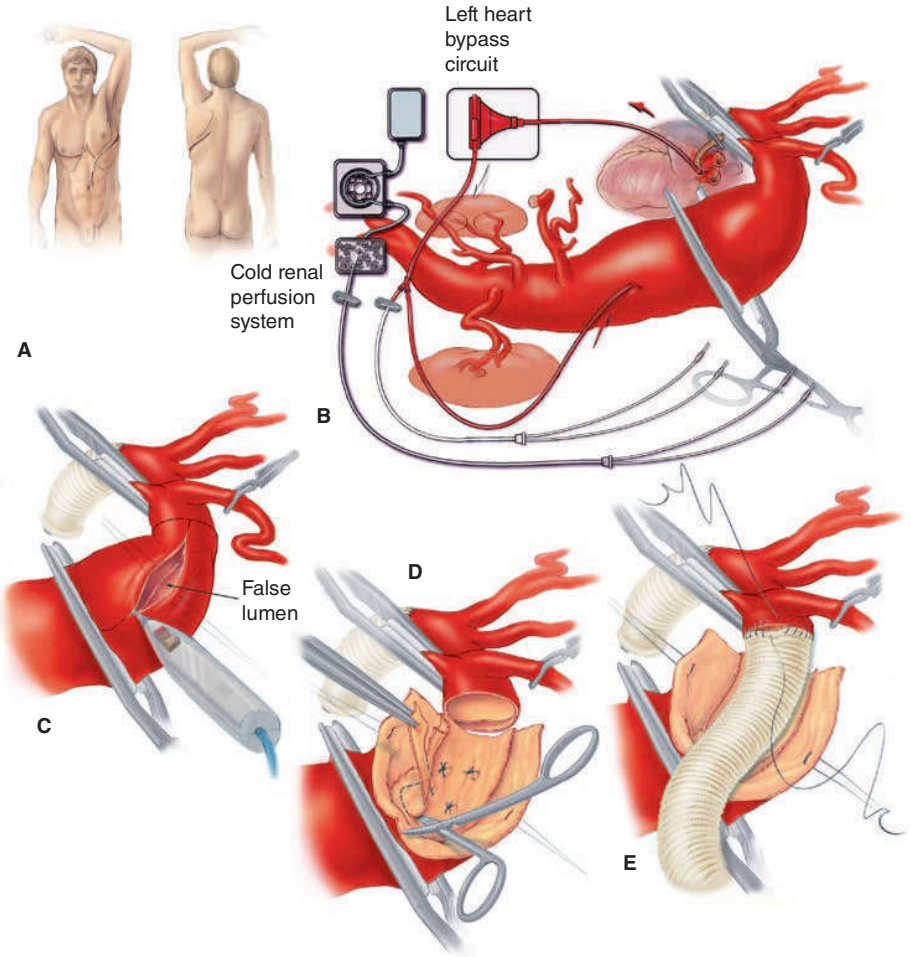


Figure 22-24. Illustration of distal aortic repair of a chronic dissection. **A.** Thoracoabdominal incision. **B.** Extent II thoracoabdominal aortic aneurysm resulting from chronic aortic dissection. The patient has previously undergone composite valve graft replacement of the aortic root and ascending aorta. After left heart bypass is initiated, the proximal portion of the aneurysm is isolated by placing clamps on the left subclavian artery, between the left common carotid and left subclavian arteries, and across the middle descending thoracic aorta. **C.** The isolated segment of aorta is opened by using electrocautery. **D.** The dissecting membrane is excised, and bleeding intercostal arteries are oversewn. The aorta is prepared for proximal anastomosis by transecting it distal to the proximal clamp and separating this portion from the esophagus (*not shown*). **E.** The proximal anastomosis between the aorta and an appropriately sized polyester graft is completed with continuous polypropylene suture. **F.** After left heart bypass has been stopped and the distal aortic cannula has been removed, the proximal clamp is repositioned onto the graft, the other two clamps are removed, and the remainder of the aneurysm is opened. **G.** The rest of the dissecting membrane is excised, and the openings to the celiac, superior mesenteric, and renal arteries are identified. **H.** Selective visceral perfusion with oxygenated blood from the bypass circuit is delivered through balloon perfusion catheters placed in the celiac and superior mesenteric arterial ostia. Cold crystalloid is delivered to the renal arteries. The critical intercostal arteries are reattached to an opening cut in the graft. **I.** To minimize spinal cord ischemia, the proximal clamp is repositioned distal to the intercostal reattachment site. A second oval opening is fashioned in the graft adjacent to the visceral vessels. Selective perfusion of the visceral arteries continues during their reattachment to the graft. A separate anastomosis is often required to reattach the left renal artery. **J.** After the balloon perfusion catheters are removed and the visceral anastomosis is completed, the clamp is again moved distally, restoring blood flow to the celiac, renal, and superior mesenteric arteries. The final anastomosis is created between the graft and the distal aorta. *(Reproduced with permission from Creager MA, Dzau VS, Loscalzo J:* Vascular Medicine*, 7th ed. Philadelphia, PA: Elsevier/Saunders; 2006.)*

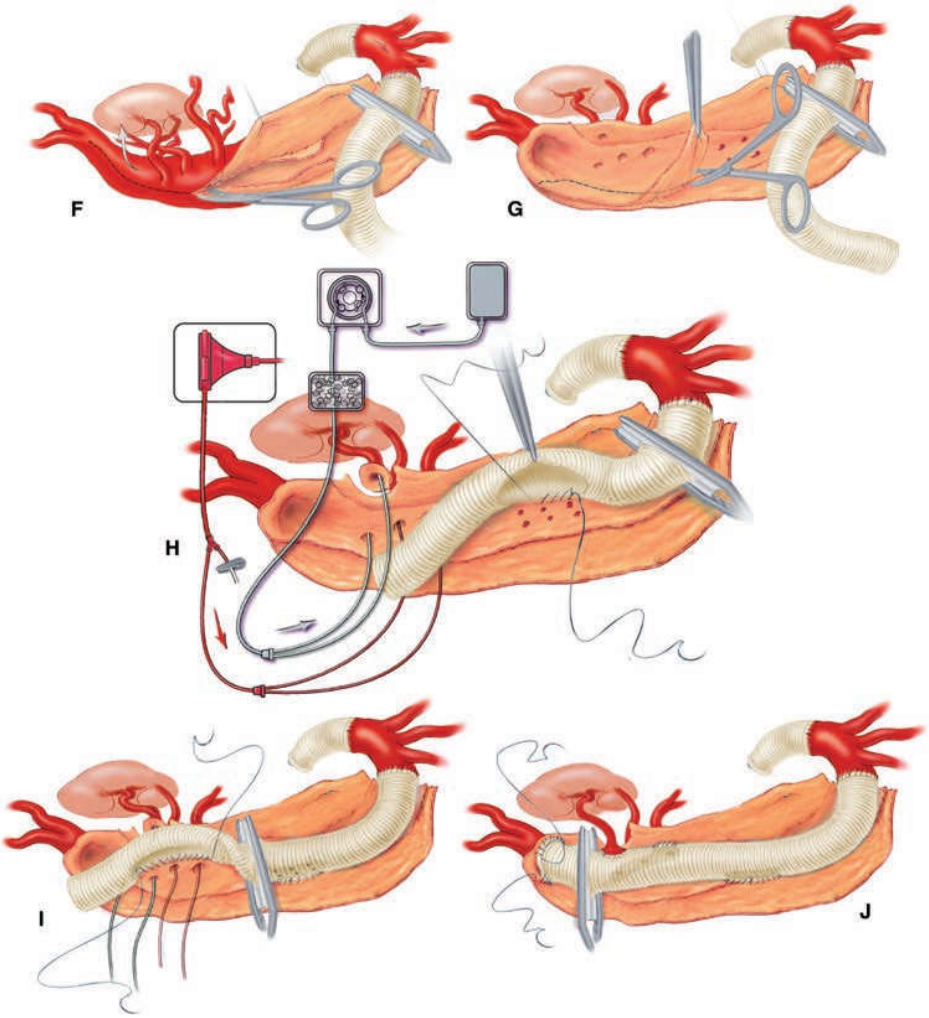


Figure 22-24. (*Continued*)

studies indicate that the enhanced risk of neurocognitive disturbances in ascending repairs using circulatory arrest are not offset by lower rates of early mortality.[227,228] Regarding extended proximal repair, reported early mortality rates after traditional stage 1 elephant trunk repairs (primarily using island reattachment strategies) range from 2.3 to 13.9%.[229-233]

Contemporary mortality rates for extensive proximal aortic repair have improved as new strategies and modified adjuncts have been adopted. For example, by adopting contemporary approaches, we have reduced early mortality for stage 1 elephant trunk repairs from 12% to 2% in our patients.[86,230] Similarly, in a report by Kazui and colleagues[234] covering 20 years of experience and 472 consecutive patients who underwent aortic arch repair with selective ACP, operative mortality was 16.0% for early repairs and 4.1% for more recent repairs. Other contemporary reports of the use of techniques such as moderate hypothermia and Y-graft approaches[235-238] indicate similarly improved outcomes; early mortality ranges from 1% to 7%, stroke rates range from 1% to 6%, and no cases of paraplegia are reported. Although paraplegia has traditionally been an unusual and infrequent complication of aortic arch repair, it has been reported as a complication of "long" elephant trunk approaches[239] and frozen elephant trunk approaches.[240]

Because of the heterogeneity of hybrid arch approaches and the tendency to use these approaches in high-risk patients, results of hybrid arch repair are difficult to interpret. In a recent report from our group,[65] among 319 consecutive patients who underwent total arch replacement in the last 8.5 years, 274 patients had traditional open repair and 45 patients had hybrid zone 0 exclusion repairs. The rate of permanent adverse outcome (death, persistent neurologic deficit at discharge, or persistent hemodialysis at discharge) was not significantly different between the two groups. A higher overall stroke rate was noticed in the hybrid group, reinforcing the importance of catheter skills and careful wire manipulation. A meta-analysis conducted by Koullias and Wheatley[241] of data from 15 studies with 463 patients found an average 30-day mortality rate of 8.3%; stroke, 4.4%; paraplegia, 3.9%; and endoleak, 9.2%. Of note, relatively few repairs (30%) were performed "off-pump," and the majority of repairs used cardiopulmonary bypass or hypothermic circulatory arrest. Additionally, several reports of small series have documented increased risk of acute retrograde aortic dissection during hybrid arch repairs; rates range from 0% to 7.5%, and these patients face significant mortality risk (ranging from 33% to 100%) should this occur.[108,109,242-244]

Treatment of Acute Ascending Aortic Dissection

The International Registry of Acute Aortic Dissection (IRAD) provides the most comprehensive data on contemporary outcomes in patients with acute aortic dissection. This registry was established in 1996 and has accumulated data from >7000 patients treated for acute aortic dissection at 51 centers in 12 countries. An IRAD analysis of data from 776 patients who underwent surgical repair of acute ascending aortic dissection revealed an in-hospital mortality rate of 23.8%.[245] The investigators identified several preoperative predictors of early mortality, including age >70 years, previous cardiac surgery, hypotension or shock at presentation, abrupt onset of symptoms, migrating pain, cardiac tamponade, preoperative renal failure, pulse deficit, and evidence of myocardial ischemia or infarction on ECG.[245,246] In a report from IRAD, in-hospital mortality after surgical treatment had decreased from 25% in 1995 to 18% in 2013.[247] The German Registry for Acute Aortic Dissection (GERAADA) has collected data on more than 3300 patients from 56 centers since 2006. [248] In a report of 1436 patients with acute proximal dissection that was surgically repaired using hypothermic circulatory arrest with or without unilateral and bilateral ACP, the early mortality rates ranged from 13.9% to 19.4%; the 628 patients with unilateral ACP had the lowest rate of early death.[249] Operative mortality reported by North American centers varies from 5% to 17%; improvements in outcomes may be related to the implementation of protocol-based management and the assembly of thoracic aortic teams.[203]

Repair of Distal Aortic Aneurysms

Endovascular Repair of Descending Thoracic Aortic Aneurysms. In the earliest series of endovascular repairs of descending thoracic aortic aneurysms, mortality and morbidity were difficult to assess. Most of the reported series were small and included a large proportion of high-risk patients with substantial comorbidity.[250,251] Subsequent evidence from pivotal, nonrandomized trials that compared patients who underwent endograft exclusion with historical or concurrent patients who underwent open repair[252-254] showed that the stent graft groups had significantly less morbidity and early mortality than the open repair groups, although in two of the trials, a nonsignificant between-group difference was observed in the rate of stroke.[252,254] Five-year comparative data show that the two groups differed significantly in their aneurysm-related mortality rates (2.8% for endovascular patients and 11.7% for open repair patients) but not in their rates of all-cause mortality (which were 32% and 31%, respectively).[255] Additional pivotal trial 5-year outcomes[256] indicate the growing disparity between aneurysm-related (96.1%) and all-cause survival (58.5%) in patients with endovascular repair, leading some to comment on the possible futility of repair in many patients.[257] Among 8967 patients identified in the National Inpatient Sample database (8255 with open repair and 712 with endovascular repair), the odds of death were 46% lower among patients who underwent endovascular repair rather than open repair.[56] The endovascular repair group also had lower odds of postoperative neurologic, cardiac, and respiratory complications.

Open Repair of Descending Thoracic and Thoracoabdominal Aortic Aneurysms. Contemporary results of open repairs of descending thoracic aortic aneurysms, including those performed in select patients with chronic dissection, indicate that early mortality rates range from 4.1% to 8.0%, renal failure rates range from to 4.2% to 7.5%, and paraplegia rates range from 2.3% to 5.7%; stroke rates are generally lower, ranging from 1.8% to 2.1%.[258-260] In our series, although the risk of paraplegia increased with the extent of repair, the risk of mortality was greatest for those undergoing repair of the proximal two thirds of the descending aorta.[258] As expected, stroke rates after distal aortic repairs were highest when the clamp site was near the left subclavian artery.

Contemporary series of open thoracoabdominal aortic repairs show acceptable survival. Reported outcome rates range from 5% to 12% for early mortality, 3.8% to 9.5% for paraplegia, 1.7% to 5.2% for stroke, and 6% to 12% for renal complications.[261-265] Many of these series summarize 10 to 20 years of surgical experience,[262-265] although some present a shorter but more contemporary experience.[261] Even for complex thoracoabdominal aortic repairs, such as stage 2 elephant trunk repairs, several centers report acceptable early mortality rates ranging from 0% to 10%.[229-233] Worse outcomes are also documented, as in a statewide, nonfederal analysis of data from 1010 patients whose early mortality rate was 25%. Of note, 40% of these patients were treated at centers averaging only one thoracoabdominal aortic aneurysm repair per year.[266] Cowan and colleagues,[267] who examined the influence of familiarity with the procedure on rates of mortality and morbidity after thoracoabdominal aortic aneurysm repair, reported that patients treated at low-volume centers fared less well. Replacing the entire thoracoabdominal aorta (i.e., performing an extent II repair) carries the highest risk of death, bleeding, renal failure, and paraplegia.[118,262,263] Early survival has been estimated at 79% at 2 years,[268] and mid-term survival has been estimated at 63% at 5 years.[265] In our recent report of 3309 repairs,[269] the overall mortality rate was 7.5%, and the rate of operative death was higher in extent II and III repairs than in extent I and IV. Permanent paraplegia and paraparesis occurred in 2.9% and 2.4% of patients, respectively, and the incidence of paraplegia in patients 50 years of age or younger was only 1.1%. Estimated survival after repair was 84% ± 1% at 1 year, 64% ± 1% at 5 years, 37% ± 1% at 10 years, and 18% ± 1% at 15 years.

Treatment of Descending Thoracic Aortic Dissection

Nonoperative Management. The in-hospital mortality rate is 8.7% for patients with acute descending aortic dissection who receive nonoperative treatment[247]; however, when IRAD stratified patients according to clinical presentation, the mortality rate for patients with uncomplicated dissection was less than 4%, whereas the mortality rate for patients with complicated dissection was more than 20%.[174,270] The primary causes of death during nonoperative management are rupture, malperfusion, and cardiac failure. Risk factors associated with treatment failure—defined as death or need for surgery—include an enlarged aorta, persistent hypertension despite maximal treatment, oliguria, and peripheral ischemia. Among patients who receive nonoperative treatment for descending aortic dissection and who survive the acute period, approximately 90% remain alive 1 year later, and approximately 76% are alive 3 years later.[271]

Endovascular Treatment. For patients with complicated acute descending thoracic aortic dissection, including rupture and malperfusion of the visceral or renal arteries, an endovascular approach is ideal. The Stanford group reported a 93% technical success rate for endovascular reperfusion of an ischemic bed.[272] Their experience with the use of first-generation stent grafts to treat acute complicated descending dissections

was also encouraging: Complete thrombosis of the false lumen occurred in 79% of patients. The early mortality rate was 16%, comparable to that associated with open techniques.[273] A meta-analysis of observational studies of endovascular repair, which included 248 patients with acute descending thoracic aortic dissection, found a 30-day mortality rate of 9.8%.[274] When compared with early mortality rates obtained from IRAD data,[174] this rate is substantially lower than the rate associated with open surgical treatment and is similar to the rate achieved with nonoperative management. However, patients with complicated acute descending dissection remain susceptible to late events; at 1 year, survival is approximately 70%, and reintervention is needed in about 10% of survivors.[275]

The ADSORB trial[276] focused on patients with uncomplicated acute descending thoracic aortic dissection. Patients were randomly assigned to optimal medical therapy alone (n = 31) or endovascular repair plus optimal medical therapy (n = 30).[277] The 1-year results showed aortic remodeling with false lumen thrombosis and reduced diameter in the group treated with endovascular repair.

The INSTEAD-XL trial involved 140 patients with stable, early-chronic descending thoracic aortic dissection who were randomly assigned to either endovascular repair plus optimal medical treatment or optimal medical therapy alone.[211] The eagerly anticipated 5-year data showed that endovascular repair was associated with greater survival and slower disease progression.

CONCLUSIONS

Aortic aneurysm may present as localized or extensive disease. The availability and development of adjuncts and endovascular techniques have supported the constant evolution of surgical strategies to tackle these complex problems. Repair strategies range from isolated, totally endovascular aortic repair for descending thoracic aneurysms to extensive total aortic and staged replacements with a combination of both open and endovascular techniques. Regardless of the difficulty of accurately assessing the risks associated with aortic repair, surgical repair of the thoracoabdominal aorta clearly remains the most challenging aortic repair in terms of mortality and morbidity. Accordingly, replacing the entire thoracoabdominal aorta (i.e., performing an extent II repair) carries the highest risk of death, renal failure, and paraplegia.[69,70,261,263,269]

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