Aortic Diseases

Ascending and Arch Aorta
Pathology, Natural History, and Treatment

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Aortic aneurysms are the 13th-leading cause of mortality in the United States. The incidence of thoracic aortic aneurysms (TAA) is increasing with improvements in screening, as well as advances in imaging. Replacement of the ascending aorta accounts for the majority of thoracic aortic procedures. TAAs are more frequently present in men and typically occur in the 50- to 70-year age range. Disease processes affecting the ascending and arch aorta include degenerative aneurysms and aneurysms associated with connective tissue disease, as well as acute aortic dissection and its variants of intramural hematoma and penetrating ulcer. Syphilitic aneurysms, once the predominant cause of ascending aortic aneurysms, are exceedingly rare today. In the present review, we will discuss these pathological conditions as well as operative techniques and outcomes after medical and operative therapy.

The Spectrum of Thoracic Aortic Pathology
Degenerative Aneurysms

Degenerative aneurysms comprise the majority of those seen in the ascending aorta and have a specific pathological profile. Whereas the elastin content in the ascending aorta is high, that seen in ascending aortic aneurysms is significantly reduced. In addition, the media of the aneurysm displays a loss of smooth muscle cells and fragmentation of the elastic fibers from a process known as cystic medial degeneration. Although this process is seen normally as a consequence of aging, it is accelerated in some and results in the phenotypic expression of an ascending aortic aneurysm. Recent studies have focused on differences in ascending aneurysm pathogenesis for patients with bicuspid and tricuspid aortic valves, with the former suggested as a more-aggressive variant.

Marfan Syndrome

Marfan syndrome is the most common inherited connective tissue disease, with an incidence of 1 in 10 000. The basic genetic defect is a mutation of the gene for fibrillin-1, an essential protein of microfibrils. The phenotypic manifestation is that of disorganized elastic fibers, premature cystic medial degeneration, and a resulting complex of ocular, musculoskeletal, central nervous system, and cardiovascular abnormalities. The predominant cause of mortality is from rupture or dissection of the dilated aortic root, which is seen in 75% of patients with Marfan syndrome.

In a landmark study, Gott and associates described a multicenter observational analysis of the effects of early operative intervention on the root and ascending aorta in patients with Marfan syndrome. In this report, elective aortic root replacement was associated with a 1.5% early mortality rate, and this contrasted with a rate of 11.7% in those undergoing emergency repair. This focus on early intervention for aortic pathology, as well as advances in imaging, has extended the median life expectancy of a patient with Marfan syndrome from 42 years in 1972 to 71 years in 2000. Although the therapy for Marfan syndrome involves focusing specifically on the aortic root, any portion of the aorta is at risk for rupture or dissection as a consequence of its weaker nature.

Type A Aortic Dissection and Its Variants

Type A aortic dissection (AD), defined here as the presence of dissection proximal to the left subclavian artery, represents a true cardiac surgical emergency. Its mortality if left untreated has been estimated from classical studies at 1% per hour for the first 48 hours and can result in a mortality rate exceeding 80% in the first month. More recent studies evaluating the effects of maximal anti-impulse therapy in nonoperative candidates suggest that the mortality rate with maximal medical management has receded but still exceeds that seen with contemporary reports on operative management.

The pathogenesis of AD remains debated, with 2 prevailing hypotheses. The first presumes that the initiating event is a tear in the intima (primary tear), which then allows blood to flow into the aortic wall media creating the false lumen. The alternative hypothesis suggests that the initial event is ruptured vasa vasorum creating intramural hematoma. This hematoma results in increased wall stress during diastole and allows for intimal disruption. Although the initiating events remain debatable, the end result remains lethal, with ultimate propagation of a false channel along a predictable spiral course from the right anterior ascending aorta, then curving posteriorly into the arch and down the left aspect of the descending and thoracoabdominal aorta. Risk factors for
Aortic dissection include those contributing to an increased intraluminal pressure (eg, hypertension, hypervolemia) or those diminishing aortic wall strength (eg, connective tissue disease). Presenting symptoms include severe tearing chest or back pain; however, manifestation as a consequence of associated branch vessel compromise (eg, myocardial ischemia, severe abdominal or lower extremity pain, lower extremity paralysis, and stroke) can also occur.9

It is the latter manifestation that presents the highest risk cohort. In this group with branch vessel ischemia, malperfusion can exist by 2 different mechanisms. Two predominant mechanisms exist by which malperfusion can occur, and these have previously been defined by our group.10 In static obstruction, the dissection flap enters the branch vessel lumen without an adequate reentry tear (or a diminutive reentry tear) within the course of that artery. The compromised true lumen of that artery then becomes the sole source of inflow into that end organ. In contrast, in dynamic obstruction, the mobile aortic dissection flap intermittently covers the orifice of the branch vessel during the cardiac cycle, thus impeding arterial inflow into the end organ. The optimal timing of aortic repair in this subset of patients with acute dissection is debated. Although immediate repair with resection of the primary entry tear may eliminate dynamic obstruction, the effects of end-organ ischemia may lead to a severe reperfusion injury and its consequences. We previously suggested a strategy of delay in operation for that group with type A dissection and severe end-organ ischemia and dysfunction.9 In that group, malperfusion was relieved by a percutaneous fenestration procedure, and operative repair was undertaken after resolution of the ischemia-reperfusion injury. Others however have suggested acceptable early results with immediate operative repair.11,12 Regardless of the timing of surgery, the presence of malperfusion remains an important adverse risk factor for mortality, particularly when it involves the mesenteric or cerebral circulation.9–12

Variants of true “double barrel” AD include intramural hematoma (IMH) with or without penetrating ulcer. These variants are often associated with the elderly as well as with women.13 Analysis of a nonoperative strategy has suggested a more benign course for type A IMH when compared with that seen in true aortic dissection, particularly if not associated with a penetrating ulcer. However, recent studies from von Kodolitsch et al, as well as Ganaha and colleagues, emphasized the need to proceed with aortic repair to prevent the risk for progression to a true double barrel dissection or aortic rupture.14,15 The current recommendation is to proceed with aortic repair in the setting of acute type A IMH with or without penetrating ulcer.

**Natural History of Thoracic Aortic Disease**

Thoracic aortic intervention is typically undertaken in the asymptomatic setting. Symptoms typically occur in the setting of either a complication of the disease (ie, rupture or dissection) or when these complications are imminent. The importance of understanding the natural history of the disease is paramount, because the indications for intervention are typically to improve survival not quality of life. Although the natural history of thoracic aortic disease is not as well characterized as that for abdominal aortic disease, recent reports have yielded important information to assist in determining timing of operative therapy.

Recent data have suggested that growth of the ascending and arch aorta is a relatively indolent process.16,17 Previous studies have suggested a mean annual growth rate of 0.07 to 0.2 cm/y for this aortic segment. Risk factors for increased growth have included increasing age, female sex, presence of chronic obstructive pulmonary disease and hypertension, and positive family history, as well as the presence of aortic dissection. Finally, growth rates of TAA have also been shown to be dependent on initial aortic diameter, with larger aneurysms growing faster than smaller counterparts.

The importance of aortic diameter in determining risk for complications has been demonstrated in numerous studies. The normal ascending aortic diameter is 2 to 3 cm depending on patient age, size, and sex. The risk for aortic rupture, dissection, or death for the ascending aorta relative to absolute size was recently evaluated by Davies et al.18 They identified that the median aortic diameter at the time of rupture for the ascending or arch aorta was 6 cm. They also demonstrated a progressively increasing risk for rupture, dissection, or death culminating at 15.6% for aortic diameters >6 cm. With these data, the recommendation to intervene was set at 5.5 cm for ascending aortic aneurysms. Whereas the focus of this and other early studies was on determining the absolute size criteria for intervention, more recent reports have suggested that absolute size may not be the only appropriate variable. Indeed, female sex and increasing age have been associated with an increase in event rate.16 In addition, in most natural history studies, those patients presenting with larger aneurysms were often immediately sent for surgery, thus altering the follow-up available for these versus small aneurysms. In recognizing the differences in aortic diameter relative to sex and body size, the Yale group recently suggested the use of an aortic size index, where the maximum aortic diameter was referenced to body surface area.19 In this study, an indexed aortic size >4.25 cm/m² correlated with an event rate of 20% to 25%. Importantly, even patients presenting with aortic sizes <2.75 cm/m² displayed an event rate of 4%; those presenting with sizes between 2.75 and 4.25 cm/m² had event rates approaching 8%. With that data, as well as the recent work from the International Registry of Acute Aortic Dissection (IRAD) consortium demonstrating the propensity for dissection at less than the typical 5- to 5.5-cm intervention target, the importance of adjunctive medical therapy, including control of hypertension and avoidance of strenuous exercise, as well as the need for continued surveillance, is justified to prevent catastrophic complications.20

The natural history of special pathological subtypes has also been recently studied. The known association of bicuspid aortic valve with ascending aortic aneurysms has been associated with a higher risk for aortic growth (0.19 cm/y versus 0.13 cm/y in nonbicuspid).21 For those patients with aortic stenosis and bicuspid aortic valve disease, the risk for rupture, dissection, or death was higher than in those without aortic stenosis. A family history of aortic aneurysm disease is also a significant risk factor for aneurysm disease. Albornoz et al
Table. Suggested Imaging Surveillance for Patients With Thoracic Aortic Aneurysms

<table>
<thead>
<tr>
<th>Aortic Pathology</th>
<th>Additional Initial Workup</th>
<th>First Follow-Up Imaging</th>
<th>Subsequent Imaging</th>
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</thead>
<tbody>
<tr>
<td>Newly diagnosed TAA</td>
<td>Echocardiography to evaluate aortic valve structure and function</td>
<td>CT scan at 6 months</td>
<td>1) Annual CT scan if stable</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) Annual echocardiography if initial study demonstrated moderate to severe aortic stenosis or insufficiency</td>
</tr>
<tr>
<td>Rapidly growing TAA</td>
<td>1) Echocardiography</td>
<td>CT scan at 3 months unless indication for operation exists</td>
<td>1) CT scan at 6 months if stable, then annually thereafter</td>
</tr>
<tr>
<td></td>
<td>2) Right and left heart catheterization</td>
<td></td>
<td>2) CT scan every 3 months if growing further</td>
</tr>
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<td></td>
<td>3) Carotid duplex</td>
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<td></td>
<td>4) Pulmonary function testing</td>
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<td></td>
</tr>
<tr>
<td>Residual distal aortic dissection</td>
<td>None</td>
<td>CT scan 3 months postoperatively</td>
<td>Annual CT scan if stable distal aortic dimension</td>
</tr>
<tr>
<td>after repair of type A dissection</td>
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<tr>
<td>Known TAA in setting of pregnancy</td>
<td>Echocardiography</td>
<td>Six to eight weeks with repeat echocardiography</td>
<td>1) Echocardiography every 6 to 8 weeks including into first 3 postpartum months</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) CT scan postpartum, then algorithm per rapidly growing TAA</td>
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recently demonstrated that of 101 non-Marfan syndrome patients with thoracic aneurysms, 21.5% demonstrated an inherited pattern for TAA with differing penetrance and expression. Those with a familial type presented earlier and displayed higher growth rates than those with sporadic types. In addition, aneurysms frequently coexisted in other locations, including in the abdominal aorta and cerebral circulation in this patient group.22

Medical Therapy of Thoracic Aortic Aneurysms

Medical therapy for TAA has typically been associated with a dismal prognosis. McNamara and Pressler called to attention the high risk of unrepaired TAAs.23 Recent reports on medical therapy, often focused on patients with Marfan syndrome, have suggested improvements in outcomes for TAA. In a randomized prospective trial, Shores demonstrated that propranolol administration was associated with improvements in both aortic growth and 10-year survival for patients with Marfan syndrome.24 Similarly, Zierer et al suggested that the use of beta-blockers was associated with a decrease in need for aortic reoperation in patients after repair of aortic dissection.25 In an evaluation of angiotensin-converting enzyme inhibitors, Yetman and colleagues noted a decrease in aortic growth for Marfan patients receiving enalapril compared with those receiving beta-blockers.26 Finally, the recent experimental evaluation in a mouse aneurysm Marfan model suggested that losartan use prevents aneurysm formation, likely as a result of its transforming growth factor-beta antagonistic effects.27 This suggestion requires clinical validation in humans.

Additional important lifestyle changes include the avoidance of strenuous physical activity, particularly weightlifting.28 Pregnancy has been associated with a risk for occurrence of type A dissection, particularly in patients with bicuspid aortic valves or Marfan syndrome.29 As part of the cardiovascular effects of pregnancy, connective tissue changes occur, including dilation of the aorta along with an increase in cardiac output. These considerations led Immer et al to suggest that aortic enlargement ≥4 cm or an increase in aortic root diameter in patients with bicuspid aortic valve or Marfan syndrome is associated with a dramatic increase in risk of type A aortic dissection, particularly in the third trimester. They stressed the need for close surveillance and suggested the use of cesarian section, selective administration of beta-blockers including into the postpartum period, as well as prophylactic aneurysm repair if rapid enlargement is seen.29

The natural history of TAA is that of inexorable expansion. As a result, continued surveillance of small TAAs is mandatory to identify individuals who should undergo intervention and constitutes an important part of ongoing medical therapy. Our protocol for this surveillance is detailed in Table. Note should be made of the significant interobserver differences—by as much as 5 mm—as detailed by Cayne and associates.30 Therefore, diligent follow-up with standardized protocols is necessary to ameliorate the risk for rupture, dissection, or aneurysm-related death.

Operative Therapy for Ascending and Arch Aortic Disease

Preoperative Workup

Our usual preoperative workup for patients requiring resection of TAA includes obtaining coronary angiography and echocardiography to evaluate the need for concomitant cardiac procedures, as well as to assess the status of the aortic valve and root. Because of the frequent association of aneurysm disease with tobacco use, we routinely obtain pulmonary function testing with consultation from our pulmonary medical colleagues on a selective basis. Severe carotid stenosis represents a risk factor for stroke with operations for TAA, and therefore carotid duplex scanning is also routinely carried out. Finally, given the association of concomitant aneurysmal disease in noncontiguous aortic
segments, we ensure that a complete computed tomography (CT) evaluation of the aorta is performed before repair of TAA.3

Operative Approach and Outcomes
The routine approach for an ascending or arch TAA is via a median sternotomy. In patients presenting with prior sternotomy and aneurysms adherent to the posterior sternal table, we have found that the “clamshell” incision (bilateral anterior thoracosternotomy) is useful to avoid catastrophic hemorrhage on reentry into the chest. Cardiopulmonary bypass is used in all patients. Cannulation for cardiopulmonary bypass is often performed via the ascending or arch aorta in elective nondissected pathology. If the procedure is performed for ascending aortic dissection, the site of cannulation typically includes either the femoral or axillary arterial route to avoid inserting large bore cannulae into the weakened aortic wall.

Advantages of the axillary cannulation method include the ability to maintain cerebral flow while the patient is on lower-body circulatory arrest for operative procedures on the arch aorta. Recent data have suggested that the axillary method of cannulation may decrease the incidence of postoperative stroke for thoracic aortic operative procedures.31 The aorta is incised at the level of the pulmonary artery and then resected back to the sinotubular junction.

Examination of the root follows; concomitant replacement is indicated when the diameter exceeds 4.5 cm. When the root is dilated but the leaflets are normal (eg, Marfan syndrome), we and others have used valve-sparing root resections with good success.32,33 In this procedure, the native aortic sinuses are resected, a graft is then inserted, and the coronary artery buttons are then reattached. This procedure can be carried out with either a reimplantation technique (Figure 1, commissural posts inserted into the graft, which is anchored in place at the

Figure 1. Valve-sparing aortic root reconstruction (modified David procedure). A, The large root and ascending aortic aneurysm. Step 1 is the initial aortotomy to visualize the aortic valve. Step 2 identifies the lines of incision in the root marking the commissural posts and coronary artery buttons. B, Resected ascending aorta and root pathology after incisions have been made as detailed in A. Note the preserved aortic leaflets. C, Commissural posts and associated leaflets are inserted within the cylinder of the aortic graft (“reimplantation technique”). D, Commissural posts are resuspended within the aortic graft. E, Coronary buttons are reimplanted in between the commissural posts, thus completing the valve-sparing aortic root resection. As is evident from this figure, the aortic annulus is situated within the graft itself.
An aneurysm exceeding with concomitant arch replacement include the presence of an aneurysm.

The remodeling technique, this remodeling technique reconstructs the aorta by using a graft with 3 “tongue-like” projections shown below to replace the resected native aortic sinuses. The graft is then sutured to the patient’s remnant aortic sinus tissue adjacent to the aortic annulus. In these patients, the aortic annulus can continue to dilate and result in progressive aortic insufficiency. The remodeling technique may aerate this late complication, because the entire root including the annulus resides within the fixed diameter of the graft.

In the instance of a normal root diameter but the presence of aortic leaflet disease precluding repair, an aortic valve replacement with associated replacement of the ascending aorta is warranted. Whereas both techniques may result in excellent long-term outcomes, recent data suggest that the remodeling procedure may be less effective, particularly in those patients presenting with annuloaortic ectasia. In these patients, the aortic annulus can continue to dilate and result in progressive aortic insufficiency. The reimplantation procedure may atherate this late complication, because the entire root including the annulus resides within the fixed diameter of the graft.

After root reconstruction is performed, attention is focused on the need for arch replacement. Our indications for proceeding with concomitant arch replacement include the presence of an aneurysm >4.5 cm. Depending on the extent of disease, the arch can be resected in a proximal hemiarch manner or with an extended arch approach (Figure 3). The latter approach is often associated with longer circulatory arrest times (see Deep Hypothermic Circulatory Arrest below), though refinements in operative technique have reduced this important determinant of morbidity (Figure 3). With the proximal hemiarch resection, the arch vessels are left intact with the descending aorta as a roof and the remaining arch aorta is replaced. In contrast, extended arch resection can result in either removal of all arch tissue with bypass constructed to each great vessel or in reimplantation of an island of arch tissue containing the great vessel origins. Finally, in those patients presenting with concomitant descending thoracic aortic disease needing intervention, the second-stage operation can be set up with an elephant trunk procedure. In this approach, the distal anastomosis is created to the mid portion of a graft. The distal edge of this graft lies within the lumen of the distal aorta. In this manner, the second-stage operation can be performed without arch mobilization and by aortic cross-clamping distal to the previous anastomosis. A recent report by Greenberg et al suggested satisfactory early results with a second-stage endovascular option, obviating the need for another major open thoracic aortic procedure.

**Morbidity and Mortality From Ascending Aortic Operations**

Major morbidity after operations on the ascending and arch aorta reflects the pathology, extent of operation, and potential experience of the surgical team. For elective aneurysmectomy, rates of death or stroke range from 2% to 5%. The risk for morbidity increases with the need for arch resection, such that the risk for death increases to 5% to 7% with an attendant increase in the risk for stroke to 2% to 5%. The subset of patients with acute aortic dissection has a dramatic increase in risk for mortality and major morbidity. In a series of studies, the IRAD consortium has suggested that early mortality for repair of type A dissection exceeds 20%. Whereas a recent review of the US experience mirrors these results, other single-institution reports suggest that results can be improved at centers of excellence.

**Special Considerations for Operations on the Ascending and Arch Aorta**

**Deep Hypothermic Circulatory Arrest**

Reconstruction of the aortic arch requires interruption of blood flow to the branch arch vessels. The popularization of
deep hypothermic circulatory arrest (HCA) as an adjunctive technique to allow a “safe” period of cerebral ischemia by Griepp and colleagues revolutionized aortic arch repair. The premise of this approach relies on the observed reduction of metabolism noted with deep hypothermia (15 to 18°C), which in turn affords a degree of protection against cerebral anoxia. Clinical studies have confirmed the safety of this approach for an HCA time of 25 to 30 minutes for both memory and fine motor performance. For patients presenting with advancing age or HCA times over this limit, the incidence of a syndrome termed temporary neurological dysfunction increases. This postoperative phenomenon, seen in up to 25% of patients, manifests as altered mental status without associated focal deficits and typically resolves within the first 24 to 48 postoperative hours. Whereas temporary neurological dysfunction is relatively common after use of HCA, the incidence of stroke is much lower (2% to 8%) and has been correlated with advancing age as well as prolonged HCA duration. In a classic study, Svennson et al suggested that the “safe period” for HCA was <45 minutes and that times >60 minutes dramatically increased the risk for stroke and death.

Multiple modalities can be used for assessment of cerebral protection during HCA that may complement the use of core body temperature alone as an indicator of cerebral metabolic quiescence. These modalities include direct jugular venous sampling where the lack of extraction implies adequate cooling, use of electroencephalography to ensure minimal brain activity, and the use of cerebral oximetry with near-infrared spectroscopy. The latter modality is a noninvasive optical technique that has been shown to be correlated with improved outcome in the pediatric population, but data in adult populations are lacking.

More recent studies have focused on the adjunctive use of retrograde cerebral perfusion and selective antegrade cerebral perfusion. Improvements in neurological outcomes have been reported with use of retrograde cerebral perfusion, where flow is reinstituted via the superior vena cava and subsequently retrograde through the cerebral vasculature to emanate from the origins of the arch vessels. The mechanism of improvements seen after use of retrograde cerebral perfu-
sion remains unclear. Whereas some studies have suggested that this method may allow for delivery of oxygen and metabolic substrates, others have suggested that the primary mechanism may relate to cooling effects and the ability to prevent air embolism in the open aorta on resumption of cardiopulmonary bypass. In contrast to retrograde cerebral perfusion, antegrade cerebral perfusion, where the individual branch vessels are cannulated and flow is maintained in an antegrade manner, has been shown in numerous studies to allow prolonged lower-body circulatory arrest times without the associated risk for stroke.

Repair of Acute Type A Aortic Dissection
The goal of this operation is to resect the ascending aorta to eliminate the risk for intrapericardial rupture and to prevent coronary artery dissection or aortic valve insufficiency. More extended operative repair depends on the extent of dissection, the site of the primary entry tear, the presence of concomitant disease, and the experience of the surgeon.

The majority of patients who demonstrate root involvement with AD can be treated by obliteration of the false lumen by placement of Teflon felt as a neomedia and preservation of the aortic valve by the technique of re suspension. In this approach, pledgetted sutures are placed at the commissural posts from within the aorta to outside the reconstructed aortic wall and the normal geometry is restored. Recently reported long-term results suggest that the incidence of reoperation remains gratifyingly low in most instances. Root preservation is contraindicated in the presence of root aneurysm (≥4.5 cm), severe leaflet pathology not amenable to repair, or the presence of dissection into the coronary ostia. Root replacement is typically performed with a modified Bentall procedure with either a mechanical composite conduit, a composite of a bioprosthetic root and Dacron ascending aortic graft, or by a valve leaflet–sparring aortic root resection.

The presence of AD into the arch aorta mandates a determination of the need for arch resection. Typically, if AD is present into the distal ascending aorta or beyond, adjunctive use of deep hypothermic circulatory arrest is indicated with its attendant increase in morbidity and mortality. Indications for resection of the arch aorta include the presence of a primary entry tear in the arch or the presence of an arch aneurysm (≥4.5 cm). In the absence of these factors, the distal anastomosis can be constructed with the placement of Teflon felt as a neomedia, and obliteration of the false lumen. With recent studies suggesting a relatively low incidence of reoperation, this approach suffices for most patients. However, in noting the temporal improvements in both early and late survival, we and others have taken a more aggressive approach are lacking. In addition, the curvature of the arch aorta has made satisfactory exclusion of aneurysm sac blood flow more difficult and again long-term data are lacking.

Conclusions
Recent advances in both the understanding of the natural history of thoracic aortic disease, as well as evolving refinements in thoracic aortic reconstruction, have allowed the repair of this once formidable pathology to become more prevalent. The emerging field of endovascular therapy has potential to extend into the realm of ascending aortic repair, having already been used in the arch aorta. The dismal results with medical therapy coupled with a rise in incidence of TAAs make these exciting times for aortic specialists.

Disclosures
Dr Patel acknowledges modest speaking honoraria from Medtronic Inc, Edwards Lifesciences Inc, and W.L. Gore Inc. Dr Deeb reports no conflicts.

References


**Key Words:** aneurysm • aorta • arch of the aorta • ascending aorta • surgery