Cardiovascular disease is the leading cause of death in most Western societies and is increasing steadily in many developing countries. Aortic diseases constitute an emerging share of the burden. New diagnostic imaging modalities, longer life expectancy in general, longer exposure to elevated blood pressure, and the proliferation of modern noninvasive imaging modalities have all contributed to the growing awareness of acute and chronic aortic syndromes. Despite recent progress in recognition of both the epidemiological problem and diagnostic and therapeutic advances, the cardiology community and the medical community in general are far from comfortable in understanding the spectrum of aortic syndromes and defining an optimal pathway to manage aortic diseases.1–13 This comprehensive review is organized in two parts, with focus on etiology, natural history, and classification (with vascular staging) of aortic wall disease in Part I and emphasis on therapeutic management and follow-up in Part II. Both parts may help to better integrate the complexities of acute aortic syndromes.

V. Therapeutic Management

Medical Treatment

Patients with suspected acute aortic dissection should be admitted to an intensive care or monitoring unit and undergo diagnostic evaluation immediately. Pain and blood pressure control to a target systolic pressure of 110 mm Hg can be achieved using morphine sulfate and intravenous β-blockers (metoprolol, propranolol, or labetalol) or in combination with vasodilating drugs such as sodium nitroprusside or angiotensin-converting enzyme inhibitors. Intravenous verapamil or diltiazem may also be used, especially if β-blockers are contraindicated. Monotherapy with β-blocking agents may be adequate to control mild hypertension, and in concert with sodium nitroprusside at an initial dose of 0.3 μg/kg per minute, is often effective in a severe hypertensive state. In normotensive or hypotensive patients, careful evaluation for loss of blood, pericardial effusion, or heart failure (by cardiac ultrasound) is mandatory before administering volume. Patients with profound hemodynamic instability often require intubation, mechanical ventilation, and urgent bedside transesophageal echocardiography (TEE) or rapid computed tomography for confirmatory imaging. In rare cases, the external ultrasound diagnosis of cardiac tamponade may justify immediate sternotomy and surgical access to the ascending aorta to prevent circulatory arrest, shock, and ischemic brain damage. Percutaneous pericardiocentesis as a temporizing step has often failed, as it can accelerate bleeding and shock.14

Surgical Treatment

The aim of surgical therapy in proximal type A (type I, II) aortic dissection is prevention of rupture or development of pericardial effusion, which may lead to cardiac tamponade and death. Similarly, sudden onset of aortic regurgitation and coronary flow obstruction requires urgent surgical intervention with the aim of resecting the region of intimal tear in dissection limited to the ascending aorta and replacement by a composite or interposition graft (if the aortic valves are intact or resuspendable). When the dissection extends to the aortic arch or the descending aorta, resection of the entire intimal flap may not be possible or the patient may require partial or total arch replacement. The largest published experience with various methods of surgical interventions in type A dissection was recently reported from Stanford.15 This report highlights the problem of either resecting or leaving unrecognized intimal tears in the arch or descending thoracic aorta, which is seen in 20% to 30% and predisposes to later distal aortic reoperation16 (Table 1). Considering an operative mortality rate between 15% and 35% even in centers of excellence, adjunctive measures such as profound hypothermic circulatory arrest and selective retrograde perfusion of head vessels17 have been used in the surgical management of arch repair or an open distal anastomosis. Whereas the latter recently gained growing acceptance for improved outcome with a 5-year survival rate of 73±6%, profound hypothermic circulatory arrest failed to improve early complication, survival, and distal reoperation rates in patients with acute type A dissection; 30-day, 1-year, and 5-year survival estimates...
were 81±2%, 74±3%, and 63±3%, and thus not different from other techniques using propensity-matched retrospective analysis. The key to success is rapid surgery before any hemodynamic instability or deterioration.

Once the patient is on extracorporeal circulation and preferably antegrade cerebral perfusion, which is usually established after cannulation of one femoral artery and the right atrium, the aorta is mobilized to visualize the innominate artery and the aortic root. If the valve leaflets are intact, aortic valve reconstruction using David’s or Yacoub’s resuspension technique is gaining growing acceptance over valve replacement.\(^{18,19}\)

The approach to an acute type A (type I, II) dissection in a previously ectatic proximal aorta requires a different approach. In such instances, mostly in patients with the Marfan’s syndrome, a composite graft (aortic tube graft with integrated valve) is preferred with coronary reimplantation.\(^{20–22}\) Surgical allografts and xenografts are experimental because late postoperative degeneration may require reoperation on the aortic root. Valve-sparing operations are delicate endeavours in an emergency and require great surgical competence in centers with expertise in elective cases.

Technical considerations for the management of a dissected nonectatic aortic root first address the condition of the aortic valve commissures, as well as the presence and extent of the dissection in relation to the coronary ostia. If the dissection compromises the left or right ostium without disrupting the coronary vessel, the ostium can usually be preserved. An ostium completely surrounded by dissected aortic wall may be excised in button form. The dissected layers around the ostium are conjoined using tissue adhesive and over-and-over suturing before the anastomosis to a tube graft is accomplished. Bypass grafting of coronary arteries using saphenous vein segments is limited to those instances in which a small torn ostium precludes reconstruction.

### The Aortic Arch in Acute Type A (Type I and II) Dissection

Treatment of the acutely dissected aortic arch remains an unresolved issue. The discussion is centered around the question of when and to what extent the arch should be replaced. At present, there is growing consensus that any dissected arch should be explored during a short period of hypothermic circulatory arrest.\(^{15}\) In the absence of an arch tear, an open distal anastomosis of the graft and the conjoined aortic wall layers at the junction of the ascending and arch portions is justified. Arch tears occur in up to 30% of patients with acute dissection.\(^{24–26}\) If an entry tear traverses the aortic arch, the distal graft to aortic anastomosis is usually tailored to replace the arch beyond the entry-bearing portion. Whenever extensive tears are found that continue beyond the junction of the transverse and descending aortic segments, or with an acute dissection of a previously aneurysmatic arch, subtotal or total arch replacement may be required with reconnection of some or all supraaortic vessels to the graft during hypothermic circulatory arrest and antegrade head perfusion.\(^{17,27}\)

In dissecting and nondissecting aneurysms extending to the downstream aorta, an elephant trunk extension of the arch graft is an option, as first described by Borst et al.\(^{28}\) This technique greatly facilitates later procedures on the downstream aorta. Instead of performing a conventional anastomosis between the end of the graft and the descending aorta, the graft is allowed to float freely in the aortic lumen. In a later procedure, the elephant trunk section of the graft may either be connected surgically to the distal descending portion directly or extended with another tubular prosthesis, or interventionally by a customized endovascular stent-graft, which may then be anastomosed at any desired downstream level of the aorta (Figure 1).

In summary, surgery is advised without delay in acute type A (type I and II) dissection. The aim of surgery is both to prevent aortic rupture, pericardial tamponade, and death, and to relieve aortic regurgitation. Implantation of a composite graft in the ascending aorta with or without reimplantation of coronary arteries is a simple standard in a world of emerging new and sophisticated surgical options.

### Surgery in Type B (Type III) Aortic Dissection

In the current era, indications for operative treatment in patients with acute type B (type III) are limited to the prevention or relief of life-threatening complications such as
intractable pain, a rapidly expanding aortic diameter, or development of periaortic or mediastinal hematoma as signs of imminent aortic rupture, although there is growing interest in managing these scenarios by interventional stent-graft placement. Dissection of a previously aneurysmatic aorta may by most centers still be considered a surgical emergency. The onset of other complications such as malperfusion of vital aortic side branches warrants interventional therapy by stent-grafting or catheter-guided fenestration of an occlusive lamella, thus improving aortic true lumen flow. When this approach does not lead to prompt relief of symptoms, surgical intervention may still be required. At present, uncomplicated type B (type III) aortic dissections are usually treated conservatively because surgical repair has no proven superiority over medical or interventional treatment in stable patients. In complicated cases, the concept of interventional stent-graft placement is currently replacing surgical interventions in experienced centers.29–32

**Interventional Therapy by Stent-Graft Placement and/or Fenestration**

Conventional treatment of type A (type I, II) dissection consists of surgical reconstruction of the ascending aorta with restoration of antegrade flow into the true lumen; thus, interventional endovascular strategies have no clinical application except to relieve critical malperfusion before surgery by distal fenestration in type B (type III) dissection with ischemic complications. Treatment goals may also include reconstruction of the thoracic aortic segment by sealing entry tear with a Dacron-covered stent and potentially inducing aortic remodeling.29–31,33 Various scenarios of true lumen, including compression resulting in malperfusion syndrome, are amenable to endovascular management. Other examples include dynamic by intima invagination, or static collapse of the aortic true lumen, dynamic or static occlusion of one or more vital side branches, or enlarging false aneurysm due to patent proximal entry tear. Aims of treatment include reconstruction of the thoracic aortic segment containing the entry tear, induction of thrombosis of the false lumen, and remodeling of the entire aorta and reestablishing of side branch flow (Figure 2).

With surgical repair of the dissected thoracic aorta, >90% of peripheral pulse deficits can be reversed. However, patients with mesenteric or renal ischemia do not fare as well. The mortality rate of patients with renal ischemia is 50% to 70% and as high as 87% in mesenteric ischemia.34–36 The surgical mortality rates in patients with peripheral vascular ischemic complications are similar to those with mesenteric ischemia, with an 89% in-hospital mortality rate.10,37–39 Operative mortality of surgical fenestration varies from 21% to 61%, and thus percutaneous interventional management using endovascular balloon fenestration of a dissecting aortic membrane to treat mesenteric ischemia has emerged as a niche indication in such complicated cases of malperfusion.38–40

The interventional management of aortic dissection and the use of stent-grafts evolved slowly in anticipation of the risk of paraplegia from spinal artery occlusion. This complication occurs in up to 18% after surgery.39,40 However, with further technical improvement, a large series of aortic dissection cases have now been successfully treated by percutaneous stent-graft placement covering the entry tear in the descending aorta and even in the aortic arch. Recent studies have demonstrated that closure of the entry tear is essential for reconstructing the true lumen and reducing total aortic diameter. Entry tear closure promotes both thrombus formation in the false lumen and healing.29–31,40 Combined surgical and interventional procedures even for proximal dissection are on the horizon.

**Indications for Fenestration and Stent Placement**

The exact role of percutaneous fenestration and stent placement in the treatment of aortic dissection continues to evolve. There appears to be a role for stent-graft placement in the treatment of static or dynamic obstruction of aortic branch arteries because static obstruction of a branch can be overcome by placing endovascular stents across the vessel origin, and dynamic obstruction may benefit from stents in the aortic true lumen with or without additional balloon fenestration. In
classic aortic dissection, successful fenestration leaves true lumen pressure unchanged. Sometimes bare stents must be deployed in the true lumen in order to buttress the flap in a stable position remote from branch artery origins.40 In chronic dissection, where fenestration of a fibrosed dissecting membrane may result in collapse of the connection between true and false lumen, a stent may be necessary to keep the fenestration open. A rare use of fenestration is to create a reentry tear for the dead-end false lumen back into the true lumen with the aim of preventing thrombosis of the false lumen and compromise of branches fed exclusively from the false lumen or jointly from the false and true lumen; this is a concept, however, that lacks any clinical proof. Conversely, fenestration may increase the long-term risk of aortic rupture because a large reentry tear promotes flow in the false lumen and provides the basis for aneurysmal expansion of the false lumen. There is also a risk of peripheral embolism from a patent but partly thrombosed false lumen.39,40

The most effective method to exclude an enlarging and aneurysmal dilated false lumen is the sealing of the proximal entry tear with a customized stent-graft; the absence of a distal reentry tear is desirable but not a prerequisite. Adjunctive treatment by fenestration and/or ostial bare stents may help establish flow to compromised aortic branches. Compression of the true aortic lumen cranial to the main abdominal branches with distal malperfusion may also be corrected by stent-grafts that enlarge the compressed true lumen and improve blood flow.30,31,38,40 Depressurization and shrinking of the false lumen are the most beneficial results to be gained, ideally followed by thrombosis and remodeling of the entire aorta (Figure 2).

**Technique of Percutaneous Balloon Fenestration**

The technical goal in percutaneous balloon fenestration is to create a tear in the dissection flap between the true and false lumen. Fenestration is performed from the smaller (usually the true lumen) into the larger or false lumen. Most commonly, a Roesch-Uchida, Brockenborough, or Colopinto needle is used for fenestration close to the compromised side branches. After the needle and a stiff wire are advanced from the true into the false lumen at the desired location, a balloon catheter of 12 to 15 mm in diameter and 20 to 40 mm in length is used to create a transverse tear. If intravascular ultrasound is not available, a pulmonary balloon catheter can be used as a targeting object for piercing the dissecting membrane with the needle.41

**Technique of Aortic Stent-Graft Placement**

Aortic stent-grafts are primarily used to correct compression of the supplying true lumen cranial to major aortic branches and to increase distal flow. Moreover, proximal communications should be sealed to depressurize the false lumen and induce thrombosis, fibrosis transformation, and subsequent remodeling of the aorta. Stent-graft placement across the origin of the celiac, superior mesenteric, and renal arteries is strongly discouraged for empirical reasons and has not been studied extensively.

On the basis of the measurements obtained during angiography, intravascular ultrasound, TEE, computed tomography, or MRI, customized stent-grafts should be used in covering up to 18 cm of dissected aorta and the major tear. The procedure is best performed in the catheterization laboratory with the patient under general anesthesia. The femoral artery is the most popular access site and can usually accommodate a 24F stent-graft system. Using the Seldinger technique, a 260-cm stiff wire is placed over a pigtail catheter that has been navigated with a soft wire in the true lumen under both fluoroscopic and transesophageal ultrasound guidance. Carefully advanced over the stiff wire, the launching of the stent-graft is performed with blood pressure briefly lowered to 50 to 60 mm Hg by infusing sodium nitroprusside. In the moment of stent expansion, aortic pressure transiently increases, and the system may dislodge without lowering blood pressure. After deployment, brief inflation of a latex balloon may be used for improved apposition of the stent struts to the aortic wall in order to seal proximal thoracic communications and to eventually reconstruct and remodel the aorta. Both Doppler ultrasound and contrast fluoroscopy are most instrumental for documenting the immediate result or initiating adjunctive maneuvers.

A dissection that propagates into a branch artery and causes static obstruction is treated by ostial branch vessel stents. A stent is useful when a significant gradient between the branch artery and the supplying aortic lumen exists. It is important that the aortic true luminal cross section and true lumen pressures are reassessed after flow restoration in large arteries, as stenting of arterial branch stenoses can result in significant reorientation of the dissection flap with subsequent changes in the pressure of the supplying aortic lumen. Precise stent placement is crucial, and minimal stent shortening appears advantageous.42 A summary of treatment options is listed in Table 2.

**TABLE 2. Considerations for Surgical, Medical, and Interventional Therapy in Aortic Dissection**

<table>
<thead>
<tr>
<th>Surgery</th>
<th>Medical therapy</th>
<th>Interventional therapy</th>
</tr>
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<tbody>
<tr>
<td>- Treatment of choice in acute type A dissection</td>
<td>- Treatment of choice in uncomplicated type B dissection</td>
<td>- Stent-grafts to seal entry to false lumen and to enlarge compressed true lumen</td>
</tr>
<tr>
<td>- Acute type B dissection complicated by the following:</td>
<td>- Stable, isolated arch dissection</td>
<td>- Unstable type B dissection</td>
</tr>
<tr>
<td>- Retrograde extension into the ascending aorta</td>
<td>- Stable type B dissection (chronic, ≥2 weeks of onset)</td>
<td>- Malperfusion syndrome (proximal aortic stent-graft and/or distal fenestration/stenting of branch arteries)</td>
</tr>
<tr>
<td>- Dissection in MFS</td>
<td>- Stable type B dissection (under study)</td>
<td>- Stable type B dissection (under study)</td>
</tr>
<tr>
<td>- Rupture or impending rupture (eg, saccular aneurysm formation (stent-grafts?))</td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Progression with compromise of vital organs (stenting?)</td>
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Results of Interventional Therapy
With both fenestration maneuvers and bare stents in side branches, compromised flow can be restored in more than 90% (range, 72% to 100%) of vessels obstructed from aortic dissection. The average 30-day mortality rate is 10% (range, 0% to 25%), and additional surgical revascularization is rarely needed. Most patients remain asymptomatic over a mean follow-up time of about one year. Fatalities related to the interventional procedure itself are not frequent, but death may occur as a result of nonreversible ischemic changes, progression of the dissection, or complications of additional reconstructive surgical procedures on the thoracic aorta.29–32 Potential problems may arise from unpredictable hemodynamic alterations in the true and false lumen after fenestration and side branch stenting. These alterations can result in loss of previously well-perfused arteries, as well as in loss of initially salvaged side branches. Affected arteries can also be managed by endovascular techniques such as additional stent procedures.43

Recent reports suggest that percutaneous stent-graft placement in the dissected aorta is safer and produces better results than surgery for type B dissection.29–32 Paraplegia may occur after use of multiple stent-grafts but is extremely rare, especially when the stented segment does not exceed 16 cm. Results of short-term follow-up are excellent, with a 1-year survival rate of $>90\%$; tears are readapted and aortic diameters generally decrease with complete thrombosis of the false lumen. This suggests that stent placement may facilitate healing of the dissection and sometimes of the entire aorta, including abdominal segments. However, late leakages have also occurred, underlining the need for stringent follow-up. In some patients, follow-up has revealed tears that had initially been overlooked and require additional stents.

Patients can develop an inflammatory reaction after stent implantation. This may present as an elevated C-reactive protein level in concert with fever. Both signs may disappear spontaneously or with transient nonsteroidal treatment as aortic remodeling progresses.

Complications of Interventional Therapy
In view of the fact that the postprocedural mortality rate seems largely dependent on the severity and duration of ischemia before the interventional procedures (eg, in one report half of the 30-day mortalities were due to irreversible damage sustained before the endovascular treatment), timely percutaneous stent treatment seems highly desirable.13 Considering the excess mortality of acute type A dissection with malperfusion of peripheral branches, percutaneous intervention for relief of malperfusion has been suggested before surgical repair.45 This concept, however, requires further study.

VI. Long-Term Follow-Up and Treatment
The long-term approach to patients with successful initial treatment of acute aortic dissection begins with an appreciation that such patients have a systemic illness that variably predisposes their entire aorta and potentially its larger branches to dissection, aneurysm, and rupture. Systemic hypertension, advanced age, aortic size, and presence of patent false lumen are all factors that identify higher risk, as does the entire spectrum of the Marfan’s syndrome.46–48 However, all patients merit extremely aggressive medical therapy, follow-up visits, and serial imaging. It has been estimated that nearly a third of patients surviving initial treatment for acute dissection will experience dissection extension or aortic rupture or will require surgery for aortic aneurysm formation within 5 years of presentation. Furthermore, this risk is substantial in the first few months after initial therapy.

Treatment with effective $\beta$-blockade is the cornerstone of medical therapy. By lowering both blood pressure and dP/dt, $\beta$-blockers have been shown to retard aortic expansion associated with Marfan’s syndrome46 and that associated with chronic abdominal aortic aneurysms.47 Observational studies suggest similar or better benefits in aortic dissection when compared with other antihypertensive agents.49,50 Guidelines recommend progressive uptitration of dosage to achieve a blood pressure $<135/80$ mm Hg in usual patients and $<130/80$ mm Hg in those with Marfan’s syndrome.47,51,52

Serial imaging of the aorta is an essential component of long-term treatment and follow-up of patients with aortic aneurysm (before and after surgery or stent-graft placement) in Marfan’s disease and in all cases of chronic dissection. Choice of imaging modality is dependent on institutional availability and expertise, as well as extent of aortic involvement and the age of the patient. Previous recommendations suggest follow-up imaging and examination at 1, 3, 6, 9, and 12 months after discharge and annually thereafter.51,54 This aggressive strategy underlines the observation that both hypertension and aortic expansion/dissemination are common and not easily predicted in the first months after hospital discharge. Furthermore, imaging is not confined simply to the region of initial involvement because both dissection and aneurysm formation may occur anywhere along the entire length of the aorta.

Development of an ascending aortic diameter of 5 to 5.5 cm is an indication for surgical repair in patients with Marfan’s syndrome. For others, aortas exceeding 5.5 to 6 cm warrant repair, as does distal aortic expansion to $\geq 6.0$ cm in all types of patients. As with nondissecting aneurysms, rate of growth and size of the aorta are both important factors to consider when it comes to prophylactic vascular surgery. An ascending aortic aneurysm of 5.0 cm may merit urgent repair in a young patient with Marfan’s syndrome.21 Conversely, an aneurysm of 5.0 cm for 3 years in an elderly person with well-controlled blood pressure is unlikely to rupture.

Patients who have been treated with surgery and/or endovascular stent grafting warrant similar follow-up to those whose initial strategy was limited to medical treatment. Aortic expansion, dissection, and rupture both at the surgical suture site and at a distance are common in survivors of type A dissection. Meticulous attention to blood pressure control and serial imaging are just as relevant to operative survivors as for patients in early stages of aortic pathology.

Conclusions
Considering both the aging patient population in Western societies with prolonged survival despite hypertension and
the better diagnostic strategies available to more patients, the cardioiology and cardiovascular community faces an increasing incidence of acute and chronic aortic problems that desperately need to be stratified using both early biomarkers of an inflammatory and dissecting process and functional imaging of the aortic wall. At this pivotal point in time, an elevated level of awareness in clinical cardioiology and the availability of modern imaging technology should trigger the interest in diagnosing and treating the complex of acute aortic syndromes similar to previous efforts in acute coronary syndromes. Cardiologists should improve diagnostic pathways and vascular staging in acute and chronic aortic diseases, form regional referral networks and allocation systems, and utilize uniform follow-up programs. Moreover, precise definitions of pathology using clear semantics should be integrated into prospective registries of aortic diseases by a multidisciplinary team of physicians in an attempt to validate previous retrospective observations and to make the best use of evolving diagnostic and therapeutic strategies. Finally, cardiologists are in need of credible prognostic models that can support decisions for individual patient care independent of investigators, at different times, and in worldwide locations.

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References


**KEY WORDS:** aorta • aneurysm • stents • surgery • cardiovascular diseases