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TREATMENT

In general, treatment is surgical for both ruptured and unruptured SOVA, if it is associated with symptoms. Asymptomatic SOVAs, depending on the size, may also require surgery to avoid complications such as acute rupture, which can be devastating when accompanied by hemodynamic compromise and may even result in death. The SOVA also can be complicated by endocarditis or thrombus formation with central or peripheral embolization. More recently, and increasingly, transcatheter repair is being used with increasing success. 3D live imaging is an emergent and important tool that is readily available to delineate the features of SOVA, which is important for intraprocedural guidance of surgical or transcatheter repair. 
Cardiac computed tomographic angiography can be beneficial in both diagnosis as well as surgical planning. More importantly, percutaneous closure may benefit from a combined echocardiography and thoracic aortogram information. Similarly, cardiac magnetic resonance imaging may be used with appropriate protocols in the assessment of SOVA.

More recently, percutaneous closure has been successfully completed in a few series. This recent series supports percutaneous closure with good safety and efficacy in experienced hands. The procedure is especially beneficial to high-risk surgical patients with comorbidities and multiple prior sternotomies, especially if the SOVA is the sole abnormality requiring attention. Selection of patients in skilled hands makes this an attractive alternative to surgery with very good short-term and mid-term outcomes.

When left untreated, ruptured SOVAs have a high 1-year mortality, with an estimated mean survival time after diagnosis of 3.9 years. Hence, a ruptured SOVA almost always mandates surgical intervention. When an unruptured SOVA is symptomatic with heart failure, conduction system aberrations, arrhythmias, or compressive symptoms on the right ventricular inflow of the coronary arteries or RVOT, or when there is suspicion for endocarditis, these are also reasons for surgical intervention. Asymptomatic unruptured SOVA may be monitored closely. In the event of symptoms or significant aortic valvular regurgitation or evidence that rapid growth ensues or severe enlargement occurs with or without fistulous communication, surgery may be indicated with similar criteria used for ascending aortic aneurysms. There are few data to support this approach, but it provides a framework for clinical decision making. If during follow-up of these patients, rupture or compressive symptoms or infections supervene, then surgical intervention is again recommended. The operative mortality rate is low at less than 1% in uncomplicated SOVA; however, in the presence of endocarditis or acute hemodynamic collapse, the mortality rate may be higher. Long-term survival is excellent with surgery for SOVA repair, with 5- to 10-year survival between 82% and 97%.

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1. Hope J: A treatise on the diseases of the heart and great vessels, and on the affections which may be mistaken for them. Comprising the author's view of the physiology of the heart's actions and sounds as demonstrated by his experiments on motions and sounds in 1830 and on the sounds in 1834-1835. Vol 8, ed 3, London, 1839, Churchill, p 639.
The spectrum of acute aortic syndrome. Note that although a primary intimal tear, a rupture of vasa vasorum or an ulceration of an atherosclerotic plaque may initiate the cascade, preexisting abnormalities in the aortic wall in the setting of hypertension, CTDs, or trauma facilitate the development of an acute aortic syndrome. CTDs: Connective tissue disorders (such as Marfan, Loeys-Dietz, Ehlers-Danlos type 4, Turner syndrome, bicuspid valve aortopathy).

HISTORY

Elements of what we now refer to as acute aortic syndrome have been observed as early as 1555 by the European anatomist Andreas Vesalius (1514-1564), who described antemortem a traumatic abdominal aortic aneurysm in a man who had fallen from a horse; the patient’s autopsy in 1557 by Vesalius’s colleagues confirmed the diagnosis. An intimal tear, the hallmark of aortic dissection, might have been first noted on autopsy by the German anatomist Daniel Sennert (1572-1637), Latinized to Sennertus) and published posthumously in 1650. The British royal physician Frank Nichols (1699-1778) provided the first unequivocal description of aortic dissection on autopsy of the English king George II, who had died in 1760 of an ascending aortic dissection after straining on a commode. Nichols described the aortic pathology as “a transverse fissure” of the aortic trunk.

The term “dissection” appears to have been first applied to blood vessels and the aorta in 1802 by the Swiss surgeon Jean Pierre Maunoir (1768-1861). A few years later, in 1826, the French physician René Laennec (1781-1826) introduced the term dissecting aneurysm (anévrysm dissecant). Until the introduction of aortography in 1929 by the Portuguese physician Reynaldo dos Santos (1876-1970), aortic dissection was exclusively a postmortem diagnosis. Aortography remained the primary means of diagnosing aortic dissection until the introduction of the modern imaging techniques of echocardiography, computed tomography (CT), and magnetic resonance imaging in the second half of the 20th century. The first successful surgical repair of a dissection in the descending aorta was reported in 1955 by Michael DeBakey (1908-2008), an American surgeon of Lebanese origin, and his colleagues. The first successful surgical repair of an ascending aorta dissection was reported in 1962 by the American surgeons Frank Spencer and Hu Blake.

As further discussed later, DeBakey and Stanford are the two major classifications of aortic dissection. DeBakey proposed his classification in 1966; the alternative classification was published in 1970 by researchers from Stanford University in California. Because aortic dissection is a rare diagnosis and the number of patients seen at any one hospital is small, the International Registry of Acute Aortic Dissection (IRAD) was established in 1996 to pool data from leading centers in North America, Europe, and Asia. At the turn of the 21st century, methods for percutaneous endovascular repair of aortic dissection are being developed.

CLASSIFICATIONS OF AORTIC DISSECTION

Classifications of aortic dissection take into account both temporal and special aspects.

Temporal

Dissections that are diagnosed within the first 2 weeks of presentation are termed acute; once the 2-week mark is passed, they are referred to as chronic dissections.

Spatial

Stanford and DeBakey are the two most commonly used spatial classifications (Fig. 159.2). In Stanford classification, all dissections are either type A or type B. Any involvement of the ascending aorta...
classifies the dissection as type A irrespective of whether the dissection is contained within the ascending aorta or extends further distally into the aortic arch, descending thoracic aorta, abdominal aorta, and beyond. Type B dissections are confined to the descending aorta.

The DeBakey classification is more detailed than the Stanford one. Dissections contained within the ascending aorta are termed DeBakey II, whereas those extending any length distally from the ascending aorta are classified as DeBakey I. Thus DeBakey types I and II correspond to Stanford type A. Dissections limited to the descending aorta are labeled DeBakey III; this term is thus equivalent to Stanford type B. DeBakey III dissections that are confined to the descending thoracic aorta are labeled as IIIa; those extending into the abdominal aorta are called IIIb.

**EPIDEMIOLOGY**

Aortic dissection is the most common form of catastrophic aortic disease and comprises approximately two thirds to three fourths of all AAS cases. The overall incidence of aortic dissection is low and is estimated at 0.5 to 4.0 annual cases per 100,000 individuals. Thus there are only a few thousand new aortic dissection cases diagnosed worldwide each year. Men are approximately twice as likely as women to develop aortic dissection. Aortic dissections originate in the ascending aorta much more commonly than in the descending aorta; in the initial IRAD database, Stanford type A comprised approximately two thirds and Stanford type B approximately one third of all aortic dissections. The prevalence of aortic dissection has a bimodal distribution with one cluster in younger patients (around 40 years of age) and the other in older patients (around 60 years of age). In younger patients, connective tissue disorders (such as Marfan, Loeys-Dietz, Ehlers-Danlos type 4, Turner syndrome, and bicuspid valve aortopathy) are the predominant risk factor, whereas hypertension is particularly prevalent among older patients with aortic dissection.

Risk factors mediate the pathogenesis of aortic dissection either by triggering initial events (intimal tear or vasa vasorum rupture) or by promoting chronic medial degeneration that facilitates subsequent dissection.

Systemic hypertension promotes both intimal tear formation and chronic medial denegation. Hypertension is the most commonly observed risk factor in patients with aortic dissection. It is present in about three fourths of patients with aortic dissection. Dissections in the setting of cocaine use are at least in part related to systemic hypertension. Interestingly, cocaine use is associated preferentially with type B dissections.

Certain inherited connective tissue disorders are the strongest risk factor for the development of aortic dissections, especially type A. Except for Turner syndrome, these disorders tend to have an autosomal dominant pattern of inheritance. They include Marfan syndrome (due to mutations in the fibrillin gene), Loeys-Dietz disease.
For unknown reasons, pregnancy is a risk factor for aortic dissection, particularly in Marfan syndrome and bicuspid aortic valve aortopathy. About half of all aortic dissections in women younger than 40 years occur during pregnancy, especially during the last trimester and in the early postpartum period. Occasionally, aortic dissection is iatrogenic in origin following either aortic cannulation (as during arteriography or insertion of intra-aortic balloon pumps) or surgery (primarily as a result of aortic valve surgery). Atherosclerosis, although implicated in many other forms of aortic disease, is generally considered not to be a direct risk factor for aortic dissection unless associated with a penetrating atherosclerotic ulcer.

**PATHOPHYSIOLOGY**

**Basic Features**

Aortic dissection occurs when the normal intraluminal blood flow in the aorta gains access to the aortic media and cleaves it longitudinally in either antegrade or retrograde direction. The cleavage creates an intimomedial flap that separates the abnormal false lumen from the normal aortic lumen. Disruption of the medial change that predisposes to aortopathy presents with severe, often migratory chest pain that is tearing in character. The diagnosis of aortic dissection requires a high index of suspicion given the low prevalence of the disease. Classically, aortic dissection enters a chronic phase in which the false lumen either undergoes thrombosis or remains permanently patent. Thrombosis obliterates the true lumen and allows for reestablishment of physiologic flow confined to the true lumen. False-lumen thrombosis is preceded by blood stasis, which is visualized by echocardiography as spontaneous echocardiographic contrast (“smoke”) in the false lumen. Permanent patency of the false lumen is enhanced by the presence of reentry fenestrations. The cleaved media of the chronically patent false lumen may endothelialize to give rise to the so-called double-barrel aorta. In addition, progressive weakening of the false lumen’s adventitial wall may give rise to a secondary aortic aneurysm.

**Complications**

Aortic dissection may be complicated by malperfusion syndromes, aortic regurgitation, and rupture into an adjacent cavity. Malperfusion syndromes in the territories of aortic side branches may lead to coronary and cerebral ischemia in type A dissection, or spinal, limb, and/or abdominal ischemia in type B dissections. Aortic insufficiency is a possible complication of a type A dissection. The mechanism of aortic valvular regurgitation is multifactorial including effacement of the sinotubular junction and loss of leaflet support, as well as prolapse of the intimal flap through the aortic valve into the left ventricle. Aortic dissection may rupture into the pericardial, pleural, and/or peritoneal space, leading to hemorrhagic effusions. Pericardial tamponade is believed to be the most common cause of death in type A dissection. Urgent surgical repair of the aortic dissection rather than pericardiocentesis is the treatment of choice for such a complication.

**Long-Term Changes**

If the patient survives and no surgical intervention is performed within 2 weeks of dissection onset, aortic dissection enters a chronic phase in which the false lumen either undergoes thrombosis or remains permanently patent. Thrombosis obliterates the true lumen and allows for reestablishment of physiologic flow confined to the true lumen. False-lumen thrombosis is preceded by blood stasis, which is visualized by echocardiography as spontaneous echocardiographic contrast (“smoke”) in the false lumen. Permanent patency of the false lumen is enhanced by the presence of reentry fenestrations. The cleaved media of the chronically patent false lumen may endothelialize to give rise to the so-called double-barrel aorta. In addition, progressive weakening of the false lumen’s adventitial wall may give rise to a secondary aortic aneurysm.

**DIAGNOSIS OF AORTIC DISSECTION**

The diagnosis of aortic dissection requires a high index of suspicion given the low prevalence of the disease. Classically, aortic dissection presents with severe, often migratory chest pain that is tearing or ripping in nature. Physical diagnosis is notoriously unreliable in establishing the diagnosis; pathognomonic physical findings (such as pulse deficits or focal neurological signs) occur in only one third of cases or fewer. Four imaging techniques are used in diagnosing aortic dissection: echocardiography, computed tomography (CT) (Fig. 159.3, A and B), magnetic resonance imaging (MRI) (see Fig. 159.3, C and D), and aortography. Either transthoracic echocardiography or computer tomography is the initial diagnostic test of choice for
acute dissections. Note that although one can occasionally establish the diagnosis of aortic dissection on transthoracic echocardiography (Fig. 159.4/Video 159.4, A-D), this imaging technique does not have sufficient sensitivity or specificity for the diagnosis of aortic dissection and thus should be used only as a rough screening tool.

Magnetic resonance imaging is best suited for chronic dissections. Aortography, the most invasive of the four imaging techniques, typically adds no incremental value in the diagnosis of aortic dissection compared to noninvasive techniques. However, aortography is still useful during attempts to treat or palliate aortic dissection, such as during stent placement or creation of iatrogenic fenestrations in the intimomedial tear.

**ECHOCARDIOGRAPHY IN AORTIC DISSECTION**

The principal goal of aortic dissection imaging by echocardiography, CT, or MRI is to identify the three fundamental features of aortic dissection: basic findings, complications, and long-term changes.

Transeosophageal echocardiography (Figs. 159.5 and 159.6) is the echocardiographic test of choice for the diagnosis of aortic dissection, because transthoracic echocardiography lacks sensitivity and specificity in diagnosing this disorder. Nonetheless, transthoracic echocardiography may be invaluable in visualization of complications of aortic dissection.

On echocardiography, the dissection flap separating the true lumen from the false lumen appears as an undulating membrane parallel to the long axis of the aorta. The intimomedial flap is often easier to visualize in the short axis than in the long axis of the aorta. The linear reverberation artifact in the ascending aorta should not be mistaken for type A dissection. Similarly, the band of tissue separating a prominent azygos vein from the descending thoracic aorta should not be misinterpreted as a dissection flap (Fig. 159.7/Videos 159.7, A-C).

The true lumen expands with systole and shrinks with diastole and is often smaller than the false lumen. Because the true lumen is lined by the intima and the false lumen by the cleaved media, the presence of intimal atherosclerotic changes helps identify the true

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**Figure 159.3.** Computed tomography and magnetic resonance imaging in aortic dissection. **A** and **B,** Contrast-enhance computed tomography demonstrates an acute type B aortic dissection. Axial images in **A** show the typical appearance of a dissection flap in the descending thoracic aorta (arrow) separating the true (T) from the false (F) lumen. Note the absence of dissection in the ascending (Asc) aorta. PA, pulmonary artery. Sagittal images in **B** demonstrate the typical origin (arrow) of type B dissection just distal to the origin of the left subclavian artery (LSA). **C** and **D,** Magnetic resonance imaging demonstrates a chronic type B aortic dissection extending from the thoracic aorta into the abdominal aorta. Axial image **C** and coronal image **D** demonstrate that the right renal artery (RRA) originates from the true (T) lumen while the left renal artery (LRA) originates from the false lumen leading to hypoperfusion of the left kidney. SMA, Superior mesenteric artery.
lumen. The false lumen is also more likely to feature blood stasis, giving rise to spontaneous echo contrast (“smoke”) and thrombus formation. Microbubble contrast may help in distinguishing the true from the false lumen, as the contrast typically fills the true lumen before the false lumen (Fig. 159.8/Video 159.8).

Entry sites from the true lumen into the false lumen are best visualized by color Doppler jets extending from the true lumen into the false lumen at predilection sites (a few centimeters distal to the right coronary cusp in type A dissections, or in the descending thoracic aorta just distal to the origin of the left subclavian artery in type B dissections). Similarly, exit holes may be seen on the distal portions of the dissection flap, with color jets exiting the false lumen into the true lumen.

Complications of aortic dissection are easily visualized by standard transesophageal and transthoracic echocardiographic techniques: aortic insufficiency; segmental left ventricular wall motion in case of dissection into coronary arteries; echoluent space around the heart indicative of pericardial effusion; and extension of the dissection flap into aortic branch vessels. Even though transthoracic echocardiography is inadequate for the diagnosis of aortic dissection per se, visualization of known complications on transthoracic echocardiography is an important diagnostic clue that the patient might have aortic dissection.

Long-term changes in the false lumen start with development of spontaneous echocardiographic contrast in the false lumen, which eventually leads to clot formation and obliteration of the false lumen (Fig. 159.9/Video 159.9, A-C). Transesophageal echocardiography may also be used for serial monitoring of possible aortic aneurysm formation after aortic dissection.

The major drawback of transesophageal echocardiography is its inability to visualize the portion of the thoracic aorta around the origin of the brachiocephalic trunk; this region is a blind spot due to interposition of the trachea and the left main bronchus between the aorta and the esophagus. This region, however, can often be well visualized on suprasternal transthoracic imaging.

**THERAPY AND PROGNOSIS**

Type A aortic dissection is an absolute medical emergency requiring prompt surgical repair, as likelihood of survival decreases with each passing hour. Up to 90% of unoperated patients with type A dissection die within 3 months of presentation. For type B dissections, medical therapy on average has a lower mortality than surgical repair. Thus, medical therapy is the preferred choice in treating type B dissections unless complications develop. Percutaneous endovascular stent-graft placement is becoming an alternative to
Figure 159.5. Type A dissection on transesophageal echocardiography. A, Typical origin of the type A aortic dissection flap (arrows) just distal to the ostium of the right coronary artery (RCA). B, Dissection flap (arrow) remains in the ascending aorta during diastole and does not prolapse through the aortic valve in this patient. In contrast, C and D demonstrate dissection flaps (yellow arrows) through the aortic valve (AV) during diastole. Dissection flap prolapse is one of several mechanisms that lead to aortic regurgitation (white arrow) in type A dissection. E, Circumferential dissection flap in the ascending aorta seen in a short-axis view separating the true (T) from the false (F) lumen. LA, Left atrium; LMCA, left main coronary artery; LV, left ventricle; LVOT, left ventricular outflow tract; RV, right ventricle. (See accompanying Video 159.5, B-E.)

Figure 159.6. Type B dissection on transesophageal echocardiography (TEE). A, Color-filled true lumen gives rise to the celiac trunk. The false lumen is to the left of the true lumen and shows little flow on color Doppler. B, Type B dissection in the descending thoracic aorta. Arrow points to incomplete dissection of the media at this level. Findings of strands of media tissues still intact help identify the false lumen. C, Two secondary communications (arrows) between the true and the false lumen are shown. D, Flow in these secondary communications frequently demonstrates a to-and-fro pattern on spectral Doppler. E, Three-dimensional TEE image demonstrate a dissection flap separating the true (T) from the false (F) lumen. Note the acute angle (asterisk) between the false lumen and the dissection flap. This acute angle helps identify the false lumen. (See accompanying Video 159.6, B-E.)
Figure 159.7. Masqueraders of aortic dissection flap. A, A true type A aortic dissection flap should not be confused with the reverberation artifact in the ascending aorta. Note that the reverberation artifact is located twice as deep (2x) as the anterior aortic wall (1x) that gives rise to the reverberation artifact. B and C, The wall separating the descending aorta from a prominent azygos vein should not be mistaken for a type B aortic dissection flap. In this patient the azygos vein is unusually larger because of azygos continuation of the inferior vena cava (IVC) in the setting of congenital absence of the intrahepatic portion of the IVC. LA, Left atrium; RPA, right pulmonary artery. (See accompanying Video 159.7, A-C.)

Figure 159.8. Use of microbubble contrast in diagnosis of aortic dissection. Intravenously given microbubble echo contrast (such as perfluorane) helps distinguish the true (T) from the false (F) lumen. In this patient with a dissection flap in the descending thoracic aorta (A), the contrast agent fills the true lumen first (B) and then enters the false lumen (C). Images also show a left pleural effusion (LPE), which is often a complication of aortic dissection. (See accompanying Video 159.8.)
surgical repair of type B dissections. Medical therapy is used in all patients irrespective of whether they are operated or not. A multidrug regimen including a beta-blocker is recommended to control the systemic blood pressure and to decrease the rate of rise of systemic blood pressure.

CONCLUSIONS

Aortic dissection occurs when blood enters the aortic media and tears it longitudinally. Acute aortic dissection is an absolute medical emergency requiring prompt diagnosis and, often, urgent surgery. Transesophageal echocardiography and contrast-enhanced CT are preferred diagnostic modalities in the acute setting, whereas MRI is better suited for chronic dissections. The triad of diagnostic features of aortic dissection visualized by these imaging techniques consists of (1) basic findings (intimal flap; false lumen; true lumen; entry and reentry tears); (2) signs of complications (aortic insufficiency, malperfusion of aortic branches; pericardial or pleural effusions); and (3) long-term changes (thrombosis of the false lumen; double-barrel aorta; secondary aortic aneurysm).

Please access ExpertConsult to view the corresponding videos for this chapter.

REFERENCES