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The aorta is the largest artery in the body. It carries the blood from the heart to the branch arteries that supply the rest of the body (including the head, arms, abdominal organs, and legs). The aorta has the same dimensions as a garden hose and curves up from the heart before extending down to the waist.

The aorta is identified by 3 major sections: the ascending aorta, the descending aorta, and the abdominal aorta, as shown in Figure 1. The aortic wall has 3 layers (listed here from inside to outside): the intima, media, and adventitia. These layers are made up of connective tissue and elastic fibers, which allow the aorta to stretch from pressure produced by the flow of blood. Abnormalities of the aortic wall may lead to enlargement of the aorta (aneurysm) and tearing (dissection) of the lining of the aorta.

What Is an Aortic Dissection?

An aortic dissection is a tear in the inner layer of the aortic wall, which allows blood to enter into the wall of the aorta (Figure 2), creating a new passage for blood, known as the "false lumen." Blood flow into the false lumen can cause several problems: It can rob crucial blood from the rest of the body, it can cause the dissection to spread and affect other arteries, and it can block blood flow in the true aortic channel ("true lumen"). These problems may cause decreased blood flow to vital organs. Aortic dissection also weakens the aortic wall and may lead to rupture, which may be fatal, or to formation of a balloon-like expansion of the aorta, known as an aneurysm.

Aortic dissections are uncommon, yet they are highly lethal. If untreated, an aortic dissection can be fatal within the first 24 to 48 hours. Several risk factors are associated with aortic dissections, such as high blood pressure (hypertension), genetic disorders affecting the blood vessel wall, atherosclerosis, cocaine use, and trauma. Data show that the average age for dissection to occur is in the 60s and that two thirds of dissections occur in men. However, dissections can occur in young patients, especially those with genetic disorders that affect the aorta and aortic valve.

In the Stanford classification system, dissections occurring in the ascending aorta are classified as type A dissections. Dissections occurring in the descending aorta are classified as type B dissections. Depending on the location of the dissection, treatment will vary.

What Are the Symptoms of an Aortic Dissection?

The symptoms of acute aortic dissection may vary. Most patients complain of an abrupt onset of severe pain in the chest, back, or abdomen. Others may note shortness of breath, pain in the arms or legs, weakness, or loss of consciousness (fainting). Dissections can affect the arteries supplying the heart, resulting in a heart attack. If the dissection interrupts blood supply to the brain, the patient may suffer a stroke. An aortic dissection can mimic many other conditions, so doctors must include aortic dissection among the list of possible diagnoses to avoid missing it in patients complaining of chest pain.

How Are Dissections Diagnosed?

Physicians perform a complete physical examination, including measuring blood pressure, listening to the heart, and evaluating the pulses. Features such as a heart murmur that indicate a leaking aortic valve and abnormal

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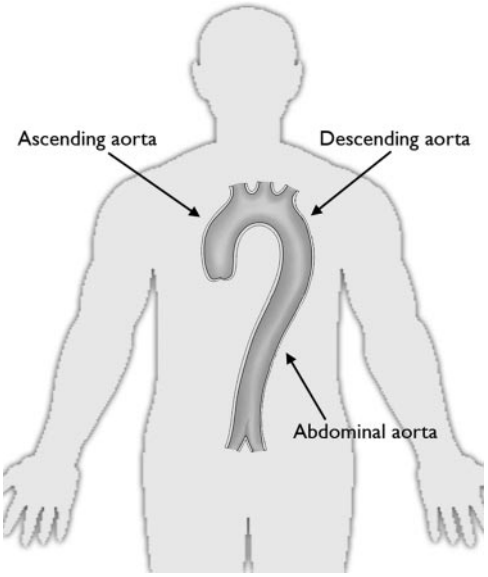

Figure 1. Divisions of the aorta including the ascending, descending, and abdominal aorta. Reproduced with permission from the International Registry of Acute Aortic Dissection (IRAD) Web site. Available at: http://www.iradonline.org.

pulses may heighten a doctor's suspicion of dissection. An electrocardiogram (ECG) may show complications of dissection, including a heart attack. The chest x-ray may show an enlarged aorta. However, both the ECG and chest x-ray may be completely normal in aortic dissection and cannot diagnose or exclude aortic dissection.

The most frequently performed tests to diagnose aortic dissection and its complications include computed tomography (CT) scan, transesophageal echocardiogram, and magnetic resonance imaging (MRI). All 3 tests are highly accurate in diagnosing aortic dissections. The specific test performed often depends on the availability and expertise of the particular hospital, as well as individual patient characteristics. CT scans require the use of intravenous dye (contrast) to visualize the true and false lumen of the aorta and branch vessel involvement. A transesophageal echocardiogram may be done at the patient's bedside and involves placing an ultrasound probe into the patient's esophagus to image the heart and aorta. Although highly accurate in diagnosing acute aortic dissection, an MRI scan takes longer than the other tests and usually is not the first test of choice.

Of note, sometimes aortic dissection may be diagnosed by a transthoracic echocardiogram, an ultrasound performed on the chest wall. Some patients require multiple different tests to confirm aortic dissection and its complications. At present, no blood tests are available to diagnose acute aortic dissection.

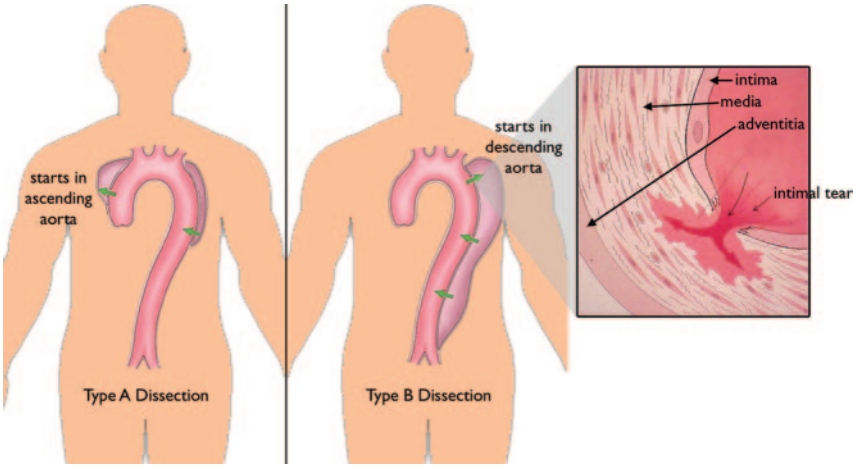

Figure 2. A, Schematic of an ascending (type A) aortic dissection, with arrows demonstrating openings or communications between the true lumen and false lumen. B, Schematic of a descending (type B) aortic dissection, with the arrows demonstrating openings or communications between the true lumen and the false lumen. Inset demonstrates tear in intimal layer. Reproduced with permission from the International Registry of Acute Aortic Dissection (IRAD) Web site. Available at: http://www.iradonline.org.

What Is the Treatment for an Aortic Dissection?

When a patient is diagnosed with an aortic dissection, the goals are to control the tear, determine whether repairing the tear would benefit the patient, and treat any complications. To control the tear, blood pressure is reduced as much as possible. Surgery is considered, and any complications are treated.

Controlling Blood Pressure

To reduce blood pressure, beta blockers (medicines that lower heart rate and blood pressure) are usually the first medications administered. If beta blockers cannot be used, calcium channel blockers such as diltiazem or verapamil are often used. Because pain can increase a patient's blood pressure, pain medication such as morphine often is needed. If blood pressure cannot be controlled with these medications, other drugs such as angiotensin converting enzyme or angiotensin receptor blocker inhibitors and/or intravenous blood pressure medications are often required.

Surgical Treatment

All patients with acute aortic dissection should be evaluated by a cardiothoracic surgeon. For patients whose dissection involves the ascending aorta

Table. Genetic Conditions Associated With Aortic Dissection

Marfan syndrome	A connective tissue disorder caused by a defect in a gene called *FBN1*. Patients with Marfan syndrome are often very tall and thin; may have long fingers, toes, arms, and legs; chest wall deformities; scoliosis; mitral valve prolapse; and eye problems (lens dislocation, myopia, or retinal detachment).
Bicuspid aortic valve	Congenital condition affecting about 1% of the general population that may occur in multiple family members. Patients with bicuspid aortic valves often have enlarged ascending aortas and are at risk for aortic dissection (5 to 10 times higher risk than the general population).
Loeys-Dietz aneurysm syndrome	Genetic disorder due to a mutation in *TGFBR1* or *TFGBR2* leading to arterial tortuosity, wide-set eyes (hypertelerism), cleft lip/palate, and aortic dissections and aneurysms involving branch vessels.
Familial thoracic aortic aneurysm/dissection	Multiple conditions involving aneurysms and dissections that occur in several family members. Currently associated with mutations in various genes including *TGFBR1* and *TGFBR2*, *FBN1, MYH11*, and *ACTA 2*.
Vascular Ehlers-Danlos syndrome (type 4)	Genetic disorder that results in a defect in the gene *COL3A1* leading to abnormal collagen synthesis. Patients with vascular Ehlers-Danlos syndrome have flexible fingers, hyperlucent skin, and varicose veins and are at risk for sudden death from spontaneous arterial dissection/rupture and intestinal or uterine rupture.

(type A dissection), immediate surgery is indicated. If the dissection involves only the descending aorta (type B), medical treatment is indicated and surgery usually is not recommended. However, if the dissection rapidly progresses, the aorta ruptures, or vital organs become threatened by lack of blood flow (malperfusion), an interventional radiologist or surgeon may use a catheter-based procedure to improve vital organ arterial perfusion, or urgent aortic surgery may be required.

Emergency surgery to repair the dissected aorta is very invasive and difficult. This typically requires a Dacron graft (a synthetic material) to replace part of the aorta to prevent blood flow into the false lumen. On average, the risk of death from acute type A aortic dissection is approximately 20%. For some patients with severe complications, the risk is much higher. Other options such as endovascular stent grafting are now being tested as alternatives to surgery in certain patients with type B dissections. In this procedure, the aorta is repaired by placing stent grafts through a leg artery into the aorta. Studies are ongoing to evaluate this approach.

Treating Complications

Because many important arteries branch from the aorta, other arteries may be affected when a dissection spreads. If the arteries that provide blood to the heart are compromised, they may need to be repaired during surgery (which could require a coronary artery bypass). The aortic valve, a 1-way valve to allow blood to flow out of the heart into the aorta, may need to be repaired or even replaced by a prosthetic valve if a dissection causes severe leaking of the valve.

After a dissection, patients will usually be required to stay in the intensive care unit so that they can be continuously monitored. Recovery from surgery usually requires 7 to 10 days. Before a patient is discharged, another CT scan or MRI is often obtained as a baseline study and to ensure that the dissection has not progressed.

Living With an Aortic Dissection

Patients are at highest risk for complications (such as aneurysm formation and recurrent dissection) during the first 2 years after an aortic dissection. Complications may begin without symptoms, so patients must be observed closely. Optimal follow-up typically consists of baseline CT scan or MRI within the first 3 months of the acute dissection and repeat imaging of the aorta at least every 6 months for the first 2 years. Over the long term, continued imaging of the aorta at least yearly is usually recommended. As many as one third of patients will subsequently require surgery on the remaining aorta because of late enlargement (aneurysm formation).

After aortic dissection, most patients require medications to control blood pressure, which ensures that stress on the aortic wall is minimized. Patients will typically be placed on beta blockers, angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, calcium channel blockers, and/or other appropriate combinations.

Lifestyle modifications are necessary to reduce the risk of long-term complications. Patients are counseled to avoid strenuous physical activity and activities that involve heavy lifting because such activity may dramatically increase the blood pressure and therefore stress on the aortic wall. Normal daily activities such as cooking, bathing, driving, and climbing stairs are not restricted. Light exercise, such as mild aerobic exercise, is usually not restricted, although contact sports are not recommended. Many patients may require a change in occupation, as sedentary jobs are often more appropriate for patients who have suffered a dissection. Lifestyle recommendations should be thoroughly discussed with a cardiologist and primary care physician.

Should I Have My Relatives Screened?

Aortic dissections have been associated with several genetic conditions, as described in the Table. Approximately 20% of all individuals with "unexplained" thoracic aortic aneurysm or dissection will have an affected first-degree relative. Screening of first-degree relatives is recommended in

each of the above conditions. These genetic conditions are associated with a weakness in the aortic wall, thus increasing the possibility that an aortic dissection may occur. Marfan syndrome, Vascular Ehlers-Danlos syndrome, Loeys-Dietz aneurysm syndrome, and familial thoracic aortic aneurysm/dissection are autosomal dominant conditions, meaning that children of an affected individual have a 50% chance of having the same genetic condition. Bicuspid aortic valves may run in families approximately 10% of the time.

First-degree relatives of patients with a family history of thoracic aortic aneurysm or dissection should be screened for aneurysms with an ultrasound from outside of the chest (transthoracic echocardiogram). For some individuals, screening with a CT scan or MRI will be necessary. In genetic conditions that could lead to aneurysms of the thoracic aorta, surgical repair is indicated once a certain size threshold is reached.

Conclusion

Aortic dissection is a life-threatening condition that must be diagnosed and treated without delay. Dissections involving the beginning portion of the aorta (type A) are treated with emergency surgery and those involving the descending aorta (type B) are usually treated with medications. After successful surgery for type A dissections, many patients may expect a relatively good short-term outlook. However, many patients require more surgery in the future and many with type B dissections will ultimately require surgery, making continued evaluation of the affected aorta with CT or MRI imaging necessary. Because aortic dissection may be related to an underlying genetic abnormality of the aorta, first-degree relatives should be screened for aortic enlargement.

Disclosures

Dr Braverman has served as an expert witness in Barlet vs Simmons et al, Bryant vs Ravitsky et al, Siluss vs Balle et al, and Brennan/Snyder vs State of New York. Dr Eagle has received research grants from W.L. Gore and Associates and has served as a consultant and on the advisory board of the National Marfan Foundation. Dr Juang reports no conflicts.

Additional Resources

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