THORACIC AORTIC ANEURYSM

Clifton Titcomb, MD
Medical Director
Hannover Life Reassurance Company of America
Denver, CO
cliff.titcomb@hlramerica.com

Executive Summary *This article discusses irregularities often seen by underwriters involving the thoracic aorta and the risk of aneurysm. Covered topics include the anatomy of the thoracic aorta, various pathologies that affect it, different types of aneurysms, common symptoms of dissection, diagnostic evaluation, potential adverse events, surgical treatments and their risks, operative mortality prognosis, and critical underwriting concerns.*

With the frequency with which asymptomatic, often minor, abnormalities are found on echocardiography, CT and MRI scanning, evaluation of the risk associated anomalies of the thoracic aorta is a major concern for underwriters. What makes this evaluation difficult is the very high mortality risk associated with unrecognized or untreated thoracic aortic aneurysms and the realization that the majority of the noted irregularities will not progress to catastrophic consequences. It is separating the wheat from the chaff in terms of risk that is hard.

Anatomy

The thoracic aorta is the main conduit from the heart to the systemic blood circulation. It has a strong elastic quality about it as is designed to function in a high pressure environment. It is this recoil ability, much like that seen with a new balloon, which allows it to absorb pressure variations and maintain a steady flow of blood to the periphery. It consists of four broad anatomic areas, the aortic root, just beyond the aortic valve, the ascending portion, the arch and the descending segment, which connects to the abdominal aorta. Along the way a number of major branch vessels originate.

The aorta is composed of three tissue layers. The innermost is the intima and includes the endothelium or inner lining of the vessel. The media is the thickest portion and that part that gives the vessel its rubbery quality. It is composed of elastic and collagen fibers as well as smooth muscle cells. The adventitia is the outermost layer and contains loose connective tissue and carries the blood vessels that supply the wall of the aorta.

Pathologies

There are several disease processes that may affect the thoracic aorta. The most important is called cystic medial degeneration. It is essentially the degeneration and loss of elastic fibers and smooth muscle cells in the middle or media layer, that portion of the vessel which gives the artery its rubbery quality, leading to a weakening of the wall. This condition especially affects the ascending aorta. It may occur as a part of the aging of a previously normal vessel and can be accelerated by the presence of hypertension. However, it may also occur prematurely in association with certain disease processes that includes; Marfan and Ehlers-Danlos syndromes and, especially, a bicuspid aortic valve. In fact, underwriters should *always* look for thoracic aorta anomalies in the presence of a bicuspid valve.

Other conditions may also lead to aortic pathology including; atherosclerosis, inflammatory disorders and trauma. Atherosclerosis can result in weakening of the wall of the vessel. While important for the arch, descending and abdominal aorta, atherosclerosis is not a significant cause for aneurysms in the root and ascending portions.

The inflammatory disorders include both infectious (syphilis) and non-infectious (vasculitis) conditions. Trauma-related abnormalities occur primarily as a consequence of damage resulting from deceleration injuries. However, it is rare to see any of these less common causes in the underwriting environment.

Types of Aneurysms

There are two distinct types of aneurysms in the thoracic aorta: dissecting and dilated. Dissecting aneurysms involve a splitting of the aortic wall which results in the creation of an artificial or false lumen. This is usually caused by one of two mechanisms. In the most common, an intimal tear allows blood, under the usual high pressure present in the aorta, to dissect into the media layer. Subsequently, the constant pounding of the systolic blood pressure drives the blood, like a wedge, splitting the vessel wall like a log, creating a false lumen. In the other mechanism, a blood vessel in the wall of the aorta, the vasa vasorum, ruptures leading to the formation of a hematoma. This hematoma in the wall of the vessel splits it, weakening it. This may then lead to the formation of an intimal tear, thus further propagating the dissection.

There are two types of dissecting aneurysms. Type A (using the Stanford classification system) involves the ascending aorta, with or without disease in the descending portion. Type B involves just the descending aorta. By definition, acute dissections are those present for less than 2 weeks and chronic ones are those lasting longer than that time frame.

Symptoms

Dissections typically present in the 5th to 7th decades of life and are more common in men. The most common symptom is pain in the chest, neck or back, which is frequently described as a tearing sensation. The symptoms are often confused with a coronary syndrome which makes diagnosis difficult. If the tear extends into or blocks branch vessels other end organ findings may ensue including; diminished pulses, coronary occlusion, stroke, spinal cord or kidney ischemia. If the damage affects the aortic valve, insufficiency may result. The most feared complication, and the major cause of death, is dissection into the pericardial sac which leads to tamponade or compression of and inability to fill the ventricle with subsequent circulatory failure. The symptoms of a dissecting aneurysm are often nonspecific making diagnosis difficult, leading to the high mortality rate associated with the condition. Survival is dependent upon an accurate and timely diagnosis and requires a high index of suspicion, especially in individuals with predisposing conditions.

Dilated aneurysms are defined as an enlargement of the aorta to more than 1.5 times the reference size in an adjacent normal segment. Dilated aortic aneurysms can be fusiform or diffuse (like a snake swallowing its prey) or saccular or localized (like a bubble on a tire). In either case, if it is large enough it can compress surrounding structures and lead to symptoms such as venous obstruction (superior vena cava syndrome), dysphagia, myocardial ischemia or hoarseness. In addition, sluggish flow in the dilated area can lead to the formation of clots which may break off and embolize. The most serious complication is rupture of the dilated area with subsequent catastrophic hemorrhage.[1,2,3]

Diagnosis

Thoracic aortic aneurysms are usually detected by imaging studies such as chest X-ray, echocardiography, CT scan or MRI. Of these the latter three are the most reliable. Several studies have documented that the normal aortic size varies with age and body surface area (BSA). What is important to remember is that the normal aortic size varies with age, sex and body size of the person. In general, older individuals, men and those with a larger body surface area have a larger expected vessel size.

Several groups have developed formulas for calculating the upper limit of normal (95th percentile) for the aortic size at the sinus of Valsalva (aortic root) and the ascending aorta. These formulas have used measurements derived from either echocardiography or CT scans and use the other factors noted above that are known to influence the vessel size. In general, athletes, particularly strength trained athletes, have larger aortic sizes than non-trained individuals, but significant enlargement in these individuals is rare. Thus, it is very important to know if the aortic size is normal or enlarged *for that person*.[4,5,6,7,8,9,10,11,12]

Potential Adverse Events

If an aneurysm is present it will likely progress over time. It follows the general medical truism that bad things get worse. The average rate of progression is 0.1 cm per year. However, this rate can vary with several factors. It is faster in the descending aorta, it is faster with conditions that weaken the wall like a bicuspid aortic valve, the presence of aortic stenosis or the presence of familial, hereditary disease and it gets faster as the aortic size gets larger. In some ways it is like a ball rolling downhill, the rate of progression accelerates as the disease progresses.[13,14,15,16,17,18,19]

Adverse events, including the development of dissection, rupture and death, are associated with the aortic size. The relationship between adverse events and vessel size holds for the measured diameter alone and for the diameter divided by the BSA (called the aortic index or AI) which corrects for the body habitus of the individual. The risk of adverse events increases dramatically to 15% or more per year at diameters of 6.0 cm or more for the ascending aorta. A similar pattern is seen with the aortic index with the highest

risk at an AI of 4.25 cm/m2 or more. The presence of chronic obstructive pulmonary disease (COPD) and pain is also associated with a higher risk aortic rupture. In contrast, the critical hinge point at which complication rates dramatically increase is higher, at 7.2 cm, for descending aortic aneurysms.[13,14,15,19,20]

However, it should be remembered that the correlation of aortic size with adverse events is greater for dilated aneurysms than for dissections. Dissections frequently occur at a smaller aortic size, especially in association with conditions that predispose the individual to cystic medial necrosis (Marfan syndrome, Ehlers Danlos, etc.).[13,14,17,21]

Other factors associated with an increased risk of rupture include: poorly controlled hypertension, the presence of symptoms, female sex and older age.

Treatment

The definitive treatment for thoracic aortic aneurysms is surgical repair, ideally before the occurrence of an adverse event. Unfortunately, an acute dissection or rupture of a dilated aneurysm frequently results in death before the individual reaches the hospital. In those patients who survive long enough to receive emergency surgery the operative mortality is high. Thus, the emphasis is on repair *before* adverse events occur.

With this in mind, the recommendation is to repair the aorta when it reaches a critical size i.e. 5.5 cm and 6.5 cm for the ascending and descending portions. For individuals with a predisposing condition such as Marfan syndrome, bicuspid aortic valve or familial history of aneurysm, the recommendation is to intervene earlier, at 4.5-5.0 cm and 6.0 cm respectively.

Other indications for surgical intervention include; any evidence of rupture, any acute dissection in the ascending aorta, a dissection of the descending aorta with major complications, rapid enlargement of an aneurysm (> 0.5-1.0 cm per year) or the development of symptoms such as pain, compression of adjacent organs, especially the trachea, esophagus or left mainstem bronchus or the development of significant aortic insufficiency.[2,19,21,22]

Prognosis

The operative mortality associated with surgical repair has improved over time but still remains high. The operative mortality is higher for emergent than for elective procedures. For those who survive the perioperative period, long-term survival studies have shown a repetitive pattern with the highest mortality risk in the first 5 years, then a decreasing relative risk over time. Thus, the longer an individual remains stable post operatively, the lower the relative risk going forward.[22,23,24,25,26,27]

Therapy differs somewhat by the location of the problem. For the ascending aorta, open surgical repair with grafts with or without valvular repair or replacement (as may be necessary with a bicuspid valve with significant aortic stenosis or insufficiency) is the only viable long-term treatment. Endovascular or stent repair is not an option and medical therapy is only used as a temporizing measure before surgical intervention. Hypertension control, especially with beta blockers, is very important postoperatively and reduces the need for reoperation.[1]

After repair of a dissecting aneurysm, the presence of a persistent false lumen is associated with a worsened outcome. The worsened prognosis results from the risk of additional dissection and a weakening of the aortic wall which may accelerate the dilation of the remaining unrepaired vessel.[1]

For dissecting aneurysms of the descending aorta medical therapy is frequently used on a long-term basis. However, as with the ascending aorta, a persistent dissection with the presence of a false lumen is associated with an ongoing risk of continuing dilation of the vessel and late complications. The definitive therapy for a descending dissection is open surgical correction but this is associated with high operative mortality and complications such as compromise of the blood flow to the spinal cord and kidneys. An alternative to open surgery in the descending aorta is the use of an endovascular stent. This approach is associated with a lower operative mortality rate and an intermediate term survival that is comparable to that with open repair. However, due to the limited long term experience they are not recommended for younger individuals. In addition, since the stents must be anchored into the surrounding non-aneurysmal aorta, they are not recommended for use in individuals with generally poor tissue quality, as seen with connective tissue diseases such as Marfan syndrome.[19]

Reoperation on a previously repaired aorta is associated with a high risk of complications and reduced survival. Several factors are associated with an increased risk of reoperation. These factors include: the presence of Marfan syndrome, a persistent dissection with the continued presence of a false lumen in the vessel wall, dilation of the residual, non-repaired aorta, absence of the use of a beta-blocker therapy post operatively and persistent, poorly controlled hypertension.[24,25,28,29,30]

Summary-Underwriting Thoracic Aneurysms

1. The aortic size should be evaluated in light of the applicant's age, sex and body habitus (BSA). Size greater than the upper limit of normal is associated with increased risk.
2. The stability of the aortic size should be evaluated over time. The longer the size has been stable the better the risk.
3. An aneurysm size progression rate of up to 0.1 cm per year is typical for an average aneurysm. A progression rate greater than this would be indicative of a higher mortality risk. The progression rate increases as the size of the aneurysm increases. It would be prudent to, at minimum, confirm that the rate of progression is absent or low.
4. Uncontrolled hypertension is a risk factor for more rapid progression and increased risk.
5. Certain medical conditions are associated with a predisposition to aneurysms and a greater rate of progression if present. These conditions include a bicuspid aortic valve, familial aneurysm conditions, Ehlers Danlos and Marfan syndrome and vasculitis/infectious causes.
6. Surgery is the definitive treatment for thoracic aortic aneurysms. For individuals without a predisposing medical condition the current size thresholds for surgical repair of the ascending and descending aorta are 5.5 cm and 6.5 cm respectively. For individuals with conditions, such as Marfan or Ehlers-Danlos syndrome, that predispose to cystic medial degeneration, the size thresholds are 4.5-5.0 cm and 6.0 cm respectively.
7. Open surgical repair is the only acceptable long-term treatment for disease of the ascending aorta.
8. Medical therapy may be used in dissecting aneurysms of the descending aorta. However, the presence of a residual false lumen is associated with a higher likelihood of progression or dilation of the vessel and a higher risk.
9. Endovascular stents are an acceptable alternative to open repair of aneurysms of the descending aorta in older individuals. They would generally not be acceptable in the ascending aorta, younger individuals and those with diseases predisposing to cystic medial degeneration such as Marfan or Ehlers-Danlos syndrome.
10. The association of coronary artery disease and/or valvular disease increases the mortality risk and should be taken into account in assessing a rating.

References

1 Patel HJ, Deeb GM, "Ascending Arch and Aorta: Pathology, Natural History and Treatment," *Circulation*, 2008; 118:188-195.

2 Novaro GM, "Diseases of the Aorta" in Carey WD, ed., Current Clinical Medicine, Second Edition, Saunders Elsevier.

3 Hiratzka JF, Bakris GL, et al., "2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/SVM Guidelines for the Diagnosis and Management of Patients with Thoracic Aortic Disease," *J Am Coll Cardiol*, 2010; 55:e27-e129.

4 Biaggi P, Mathews F, et al., "Gender, Age, and Body Surface Area are the Major Determinants of Ascending Aorta Dimensions in Subjects with Apparently Normal Echocardiograms," *J Am Soc Echocardiogr*, 2009; 22:720-725.

5 Roman MJ, Devereux RB, et al., "Two-Dimensional Echocardiographic Aortic Root Dimensions in Normal Children and Adults," *Am J Cardiol*, 1989; 64:507-512.

6 Vasan RS, Larson MG, et al., "Echocardiographic Reference Values for Aortic Root Size: The Framingham Heart Study," *J Am Soc Echocardiogr*, 1995; 8:793-800.

7 Lin FY, Devereux RB, et al., "Assessment of the Thoracic Aorta by Multidetector Computed Tomography: Age- and Sex-Specific Reference Values in Adults without Evident Cardiovascular Disease," *J Cardiovasc Computed Tomography*, 2008; 2:298-308.

8 Wolak A, Gransar H, et al., "Aortic Size Assessment by Noncontrast Cardiac Computed Tomography: Normal Limits by Age, Gender, and Body Surface Area," *J Am Coll Cardiol Img*, 2008; 1:200-209.

9 Kalsch H, Lehmann N, et al., "Body-Surface Adjusted Aortic Reference Diameters for Improved Identification of Patients with Thoracic Aortic Aneurysms: Results from the Population-Based Heinz Nixdorf Recall Study," *Int J Cardiol*, 2011; Published Online

10 Mao SS, Ahmadi N, et al., "Normal Thoracic Aorta Diameter on Cardiac Computed Tomography in Healthy Asymptomatic Adults," *Acad Radiol*, 2008; 15:827-834.

11 Hagar A, Kaemmerer H, et al., "Diameters of the Thoracic Aorta throughout Life as Measured with Helical Computed Tomography," *J Thorac Cardiovasc Surg*, 2002; 123:1060-1066.

12 D'Andrea A, Cocchia R, et al., "Aortic Dimensions in Elite Athletes," *Am J Cardiol*, 2010; 105:1629-1634.

13 Elefteriades JA, "Natural History of Thoracic Aortic Aneurysm: Indications for Surgery, and Surgical Versus Nonsurgical Risks," *Ann Thorac Surg*, 2002; 74:S1877-S1880.

14 Davies RR, Goldstein LJ, et al., "Yearly Rupture or Dissection Rates for Thoracic Aortic Aneurysms: Simple Prediction Based on Size," *Ann Thorac Surg*, 2002; 73:17-28.

15 Juvonen T, Ergin MA, et al., " Prospective Study of the Natural History of Thoracic Aortic Aneurysms," *Ann Thorac Surg*, 1997; 63:1533-1545.

16 Michelena HI, Khanna AD, et al., "Incidence of Aortic Complications in Patients with Bicuspid Aortic Valve," *JAMA*, 2011; 1104-1113.

17 Davies RR, Kaple RK, et al., "Natural History of Ascending Aortic Aneurysm in the Setting of an Unreplaced Bicuspid Aortic Valve," *Ann Thorac Surg*, 2007; 83:1338-1344.

18 Albornoz G, Coady MA, et al., "Familial Thoracic Aortic Aneurysms and Dissections – Incidence, Modes of Inheritance, and Phenotypic Patterns," *Ann Thorac Surg*, 2006; 82:1400-1406.

19 Coady MA, Ikonomidis JS, et al., "Surgical Management of Descending Thoracic Aortic Disease: Open and Endovascular Approaches, a Scientific Statement From the American Heart Association," *Circulation*, 2010;121:2780-2804.

20 Davies, RR, Gallo A, et al., "Novel Measurement of Relative Aortic Size Predicts Rupture of Thoracic Aortic Aneurysms," *Ann Thorac Surg*, 2006; 81:169-77.

21 Pape LA, Tsai TT, et al., "Aortic Diameter > 5.5 cm is Not a Good Predictor of Type A Aortic Dissection : Observations from the International Registry of Acute Aortic Dissection (IRAD)," *Circulation*, 2007; 116:1120-1127.

22 Olsson C, Thelin S, et al., "Thoracic Aortic Aneurysm and Dissection: Increasing Prevalence and Improved Outcomes Reported in a Nationwide Population-Based Study of More Than 14000 Cases from 1987-2002," *Circulation*, 2006; 114:2611-2618.

23 Olsson C, Eriksson N, et al., "Surgical and Long-Term Mortality in 2634 Consecutive Patients Operated on the Proximal Thoracic Aorta," *Eur J Cardio-Thorac Surg*, 2007; 31:963-969.

24 Zierer A, Voeller RK, et al., "Aortic Enlargement and Late Reoperation After Repair of Acute Type A Aortic Dissection," *Ann Thorac Surg*, 2007;84:479-484.

25 Tan MESH, Morshius WJ, et al., "Long-Term Results After 27 Years of Surgical Treatment of Acute Type A Aortic Dissection," *Ann Thorac Surg*, 2005; 80:523-529.

26 Meharwal ZS, Khanna SN, et al., "Ascending Aortic Aneurysm Resection: 15 Years' Experience," *Asian Cardiovasc Thorac Ann*, 2006; 14:300-305.

27 Shah PJ, Estera AL, et al., "Analysis of Ascending and Transverse Aortic Arch Repair in Octogenarians," *Ann Thorac Surg*, 2008; 86:774-779.

28 Fattouch K, Sampognaro R, et al., "Long-Term Results After Repair of Type A Acute Aortic Dissection According to False Lumen Patency," *Ann Thorac Surg*, 2009; 88:1244-1250.

29 Campbell-Lloyd AJM, Mundy J, et al., "Contemporary Results Following Surgical Repair of Acute Type A Aortic Dissection (AAAD): A Single Centre Experience," *Heart Lung Circulation*, 2010; 19:665-672.

30 Subramanian S, Roselli EE, "Thoracic Aortic Dissection: Long-Term Results of Endovascular and Open Repair," *Semin Vasc Surg*, 2009; 22:61-68.