Thoracic Aortic Dissection
An Online Continuing Education Course

PROVIDED BY: NEBRASKA METHODIST COLLEGE-
THE JOSIE HARPER CAMPUS
AN AFFILIATE OF METHODIST HEALTH SYSTEM
AND
CONTRIBUTING AUTHORS FROM THE TAD COALITION
Contributing Authors from The Methodist Health System

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John Patrick O’Gara, MD, was an adjunct faculty at Nebraska Methodist College and practiced at Methodist Hospital teaching clinical rotations for over 30 years. Dr. O’Gara graduated from the University of Nebraska in Lincoln, and the University of Nebraska College of Medicine in Omaha, followed by an internship at the Cook County Hospital in Chicago, Illinois. His general practice included several years in Los Angeles, California, a residency in neurology at University of Wisconsin- Madison, and a residency in anesthesiology at the University of Nebraska Omaha. He was the original content author when this course was conceived, which laid the groundwork for this current updated course. Dr. O’Gara is currently enjoying his retirement and is pleased to see the continued effort to educate healthcare providers on thoracic aortic dissections.
Contributing Authors from the TAD Coalition

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Loren Hiratzka, MD chaired the TAD guidelines writing committee for the AHA and ACC which also included 8 other co-sponsoring organizations, a 9th endorsing organization, with 2 additional specialty organization writing committee members. He is a practicing cardiothoracic surgeon with Cardiac, Vascular and Thoracic Surgeons, Inc., and is the medical director of cardiac surgery for TriHealth, Inc. (Bethesda North and Good Samaritan Hospitals) in Cincinnati, OH. He has served on several AHA and ACC committees and is past chair of AHA’s Council on Cardiovascular Surgery and Anesthesia, Council Operations Committee and the You’re the Cure Advocacy Ambassador Committee. He also serves on the Society of Thoracic Surgeons Workforce on National Databases.

Dianna Milewicz, MD, PhD is the President George H. W. Bush Chair in Cardiovascular Medicine at the University of Texas Medical School at Houston. She is an internist with a subspecialty in human genetics. Her research focuses on identifying genetic variations that predispose individuals to vascular diseases, such as aortic aneurysms and dissections. Milewicz is professor and director of the Division of Medical Genetics and the M.D./Ph.D. Program in the Medical School and the Graduate School of Biomedical Sciences (GSBS). She was vice chair of the Department of Internal Medicine for four years. She was named a Doris Duke Distinguished Clinical Scientist in 2002, was elected a fellow of the Executive Leadership in Academic Medicine Program in 2004, and received the Distinguished Professional Woman Award from the health science center’s Committee on the Status of Women in 2005.

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To provide an overview of thoracic aortic dissection and evidence-based practice recommendations regarding its diagnosis and treatment.
Objectives

- Review aortic anatomy, physiology, and pathophysiology.
- Recognize the importance of patient history in the diagnosis of aortic dissection.
- Describe signs and symptoms of aortic dissection through patient examination and appropriate imaging.
- Review case studies.
Credits

Nurses: 2.0 contact hour(s) awarded. Code #NE13-16 C07
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Within this text we are including aortic disease and aortic aneurysm. With reference to aortic disease, we mean the possibility of dissection of the thoracic aorta.

- **Abdominal Aortic aneurysm (AAA):** Abnormal dilation of the abdominal aorta, usually in an area of severe atherosclerosis. (Mosby, 2013)

- **Acute aortic regurgitation:** Occurs in 50% of thoracic aortic aneurysm disease patients with ascending aortic dissection. This occurs from either a circumferential tear that widens the aortic root or a disruption of the annulus by a dissecting hematoma that tears a leaflet or displaces it below the area of closure. (Mosby, 2002)

- **Aneurysm (or true aneurysm):** A localized dilatation of a wall of a blood vessel. It may be caused by atherosclerosis and hypertension, or less frequently, by trauma, infection, or a congenital weakness of the vessel wall. (Mosby, 2013)

- **Aortic dissection (AoD):** A disruption of the media layer of the aorta with bleeding within and along the wall of the aorta. Dissection may, and often does, occur without an aneurysm being present. The term “dissecting aortic aneurysm” is often used incorrectly and should be reserved only for those cases where a dissection occurs in an aneurismal aorta. (Mosby, 2002)

- **Aortic regurgitation:** The flow of blood from the aorta back into the left ventricle during diastole, resulting from a failure of the aortic valve to close completely. Also called aortic insufficiency. (Mosby, 2013)
**Abbreviations and Definitions Continued**

- **Arteriomegaly:** Diffuse arterial dilatation involving several arterial segments with an increase in diameter greater than 50% by comparison to the expected normal arterial diameter. (Mosby, 2006)

- **Ecstasia:** Suffix meaning dilatation, dilation extension or distension of an organ. (Mosby, 2013)

- **MRI:** Magnetic resonance imaging (Mosby, 2013)

- **Pseudoaneurysm (or false aneurysm):** A dilation of an artery caused by damage to one or more layers of the artery as a result of arterial trauma or rupture of a true aneurysm. (Mosby, 2013)

- **Thoracoabdominal aneurysm (TAA):** Aneurysm involving the thoracic and the abdominal aorta. (Stedman’s, 2006)

- **TEE:** Transesophageal Echocardiogram: An endoscopic/ultrasound test that provides ultrasonic imaging of the heart from a retrocardiac vantage point, thus preventing the interpose subcutaneous tissue, bony thorax, and lungs from interfering with the ultrasound. (Mosby, 2013)
Thoracic
Aortic
Dissection

The Tyler Kahle Story

http://www.tadcoalition.org/tad/4393/Tyler
Module 1: An Overview
The term “thoracic aortic disease” encompasses a broad range of degenerative, structural, acquired, genetic-based, and traumatic disease states and presentations.

Thoracic aortic diseases are usually asymptomatic and not easily detectable until an acute and often catastrophic complication occurs.

Recent data from the Centers for Disease Control and Prevention indicate that aortic aneurysm disease is the 18th most common cause of death (n=13,843) in all individuals and the 15th most common in individuals older than age 65 years (n=11,147). (National Center for Injury Prevention and Control, 2009)

Thoracic aortic aneurysm causes more deaths per year than HIV. These figures almost certainly represent underestimates of the impact of aortic diseases. Experts have suggested that 30,000 to even 60,000 deaths per year in the U.S. represents a reasonable estimate. The incidence of aortic disease is certain to increase as our population ages. (Elefteriades and Farkas, 2010)

When in the hospital with a high suspicion of thoracic aortic dissection, if the diagnosis is made promptly, and if the hospital has advanced cardiac surgical care the surgical success rate is close to 90%.
Acute thoracic aortic dissections cause sudden death in up to 40% of individuals, while survivors have an approximate 2% per hour death rate until they undergo emergent surgical repair.

If acute thoracic aortic dissection goes undiagnosed and untreated, 50% of patients will die within 48 hours.

Unfortunately, our health care system has not been able to reliably recognize this disease. Many patients with compatible symptoms — even patients at very high risk of the disease due to certain genetic factors — are misdiagnosed with tragic results.

Many triage/intake staffers miss the probability of acute thoracic aortic dissection for several reasons. These include:

- Unawareness of the emergent nature of the condition
- Variability in symptoms
- Importance of family history in the differential diagnosis
- The wide ranges in age and appearance of patients
- Underutilization of appropriate diagnostic imaging
- Increased incidence of the disease in specific patient populations
  - Including those with Marfan syndrome, Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, or bicuspid aortic valve.
- Aortic dissection is often confused with myocardial ischemia, thus leading to delayed diagnosis, misdiagnosis, or incorrectly administering antithrombotic agents (Dixon, 2011).
Familiar Faces of Thoracic Aortic Dissection

People of all walks of life have succumbed to this disease, including King George II of England. Princess Diana also died of this condition as a result of trauma to the aorta from a motor vehicle collision. Dr. Michael DeBakey, the first surgeon to repair an aortic dissection, was afflicted with an aortic dissection late in his life and had the defect repaired by his own surgical team.

The National Marfan Foundation's President and CEO, Carolyn Levering, is painfully aware of such errors in diagnosis. She wrote:

"Jonathan Larson was preparing for the off-Broadway opening of his new musical RENT when he was taken to two different hospital emergency departments over several days complaining of massive chest pains. One emergency room diagnosed him with food poisoning while the other diagnosed him with the flu. Jonathan later died alone in his apartment of a ruptured aneurysm. His height, lanky build, and indented chest bone suggested Marfan syndrome before his aortic dissection."
Likewise, actor John Ritter presented to an emergency room with chest pain and was thought to be suffering a heart attack from coronary artery disease. He was taken to the cardiac catheterization laboratory. Coronary artery disease was not found to be the cause of his pain. He died of a dissection of the aorta.

The same affliction was thought to have taken the life of his father, Tex Ritter. Thus, John’s death served as a “red flag” that other family members may be at risk for aortic aneurysms and/or dissections. After John’s death, a scan revealed that John's brother, Tom, also had a thoracic aortic aneurysm. This was safely surgically repaired.

(This information was retrieved on September 16, 2008 from http://johnritterfoundation.org.)
In this online course, you will learn that it is possible to decrease the mortality associated with acute aortic dissection. The key factors to decreasing this mortality include:

1. Improved recognition and assessment of those patients at risk of having the disease.
2. Rapid and appropriate imaging of patients to accurately make or exclude the diagnosis.
3. Emergent referral of patients with ascending aortic dissection to cardiothoracic surgery for prompt repair.
4. Education for families regarding aortic dissection as a familial disorder.
Module 1 Review Questions

Before moving on to Module 2, please answer the review questions for Module 1.
Module 2: Anatomy, Physiology, and Pathophysiology
As illustrated, the left ventricle of the heart contracts and pumps the blood through the aortic valve. The blood flows upward into the ascending aorta, the arch of the aorta, the thoracic descending aorta, and finally into the abdominal aorta. In adults, the thoracic aorta is about 3 cm in diameter at the proximal ascending aorta. The descending thoracic aorta diameter is about 2.5 cm. The abdominal aorta is approximately 1.8 cm to 2.0 cm in diameter.
The aortic wall consists of three layers:

- **Tunica Intima**
- **Tunica Media**
- **Tunica Adventitia**

The incremental changes in arterial and venous total cross-sectional areas correlate with the rapid arterial and slow venous blood flow in these respective vessels.

# Layers of the Aortic Wall

<table>
<thead>
<tr>
<th>Layer</th>
<th>Description</th>
</tr>
</thead>
</table>
| **Tunica Intima** | 1. The thin intima layer is composed of a very inner layer of endothelial cells, with a middle subendothelial connective tissue layer.  
                     2. The outer layer of the tunica intima consists of internal elastic lamina.  
                     3. This elastic lamina can expand and dilate with the rapid systolic aorta blood flow.  
                     4. This layer also constricts down in diastole, assisting in the forward movement of the blood. |
| **Tunica Media**  | 1. The middle layer of the wall of the aorta.  
                     2. This is a thickest part of the aortic wall and is composed of layers of elastic tissue and smooth muscle cells, capable of expanding and contracting. |
| **Tunica Adventitia** | 1. The outer third layer of the aorta.  
                           2. The tunica adventitia is composed of connective tissues.  
                           3. The blood supply for the aorta and nerves to the layers of the aorta also are present in this outer layer.  
                           4. These small arteries are called the vasa vasorum.  
                           5. The nerves innervating the aortic tissues are referred to as the nervi vascularis. |
Tracing of the aortic pressure curve

The incisura, as shown on the tracing, is the "notch."

This notch represents a short drop in the aortic pressure, due to a backward flow of blood in the proximal ascending aorta.

This backward flow of blood occurs immediately before closure of the aortic valve.

Damping of the pressure pulses in the proximal aorta

Typical changes in the contours of the pressure pulse as the pulse travels into the peripheral vessels.
After the aortic valve closes, the diastolic pressure falls in the ascending aortic arch of the aorta and the thoracic ascending aorta. The elastic fibers and smooth muscle fibers contract, pumping and pushing the blood flow peripherally down the abdominal aorta.

The rapid entry of blood into the thoracic aorta causes the aortic wall to expand and the aortic pressure rapidly increases to about 120 mmHg systolic pressure. At the end of systole, the aortic valve closes, as the left ventricle ceases to pump. The elastic layer allows the aortic wall to recoil down to maintain a high diastolic aortic pressure, with forward movement of the aortic blood flow.

The velocity of the aortic blood flow averages 33 cm/sec. In comparison, the blood flow of a capillary vessel is only 1/1000 of the aorta blood flow. The capillary flow is 0.3 mm/sec. The average capillary has a length of 0.3 mm to 1 mm. The blood remains in a capillary vessel for 1-3 seconds. In this brief 1-3 second period, there is the critical blood-to-cell and cell-to-blood exchange of everything important to cellular metabolism, i.e., oxygen, carbon dioxide, electrolytes, nutrient food, and cellular waste products in a reverse exchange. This cellular metabolism exchange occurs at the capillary–cellular interspace level.
In addition to the velocity of blood passing through the thoracic aorta, there is the velocity of the transmission of the aortic pressure pulse wave. This is the shock wave or the pressure wave transmission of the aortic blood flow. This shock wave is 10-15 times the velocity of the aortic blood flow. Remember, the aortic blood flow is 33 cm/sec. The aortic pressure wave or shock wave originates from the left ventricle contraction and pumping mechanism.

The wall tension of the thoracic aorta is governed by the law of La Place. This law indicates that the wall tension in a fluid-filled conducting artery/vein is the product of the fluid pressure and the radius of the artery/vein. Thus, during systole, as the left ventricle pumps and ejects blood up into the thoracic aorta, the thoracic aorta expands. The aortic blood pressure and the radius of the aorta determine the wall tension of the thoracic aorta wall.

Considering the law of La Place, along with the velocity of the aortic blood flow and the shock pressure wave, the aorta is subject to high intermittent pressures. This explains why the thoracic aorta is more likely to dilate, dissect, tear, and rupture compared to other blood vessels.
In gross appearance an aneurysm may be shaped as fusiform or saccular. Sometimes a hematoma forms in the tunica media layer and may eventually progress into a false aneurysm formation or an aneurysm rupture.

The fusiform aneurysm involves the entire circumference of a portion/segment of the aorta. This presents as a uniformly, diffusely, dilated artery. It tapers from the middle to its proximal and distal ends, where it narrows. Fusiform is derived from the Latin word, *fusus*, meaning spindle.

The saccular aneurysm involves only a portion of the circumference. This results in a bulging sac or out-pouching of a portion or side of the wall of the aorta. A saccular aneurysm is similar to a hernia-like structure extending out from the aorta wall. It is asymmetric in appearance.
An aortic dissection is caused by a circumferential or partial tear of the tunica intima layer.
Aortic Dissection

- Often occurs along the right lateral wall of the thoracic aortic aneurysm where the hydraulic shear stress is high.
- Most common location of an aneurysm is the descending thoracic aorta just below the ligamentum arteriosum.
- Initiating event is either a primary intimal tear or a secondary dissection into the tunica media.
- The pulsatile aortic flow dissects along the elastic fibers of the tunica media creating a false lumen.
  - This dissection usually propagates distally down the descending aorta, sometimes even involving and occluding major branches off the affected aorta.
- Sometimes, dissection may occur from the arch of the aorta and retrograde down the ascending thoracic aorta.
- Added to the pathology of the aorta, sometimes a secondary distal intimal disruption occurs, culminating in the re-entry of the blood from the false lumen back into the true lumen of the aorta.
The Stanford classification separates aortic dissections into two types based on the location of the dissection:

<table>
<thead>
<tr>
<th>Type A Dissections</th>
<th>Type B Dissections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Involves the ascending aorta</td>
<td>Do not involve the ascending aorta</td>
</tr>
<tr>
<td>Require surgery</td>
<td>Usually managed medically</td>
</tr>
<tr>
<td>Type A dissections of the ascending aorta may extend proximally to involve the origin of the coronary arteries or communicate with the pericardial sac. Dilation of the aortic root or disruption of the aortic valve may result in incompetence or leakage of the aortic valve.</td>
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Dr. Michael DeBakey, who performed the first successful repairs of aortic dissection, recognized three categories of aortic dissection:

<table>
<thead>
<tr>
<th>Type</th>
<th>Involvement</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I dissections</td>
<td>Involve the ascending aorta, aortic arch, and descending aorta</td>
<td>Requires surgery</td>
</tr>
<tr>
<td>Type II dissections</td>
<td>Involve only the ascending aorta</td>
<td>Requires surgery</td>
</tr>
<tr>
<td>Type III dissections</td>
<td>Involve only the descending aorta distal to the left subclavian artery</td>
<td>Usually treated medically</td>
</tr>
</tbody>
</table>
When dissection involves the origin of the coronary arteries, occlusion or compromise of these vessels can result in myocardial ischemia or infarction, mimicking traditional presentations of patients with coronary artery disease.

If the dissection extends to communicate with the pericardial sac, pericardial tamponade ensues, usually with catastrophic consequences.
Module 2 Review Questions

Before moving on to Module 3, please answer the review questions for Module 2.
Module 3: Etiology of Aortic Dissection

Photo: Courtesy of The Marfan Foundation

There are a multitude of diseases associated with aortic abnormalities. These diseases can cause aneurysms, dissections, or rupture of the thoracic or abdominal aorta. Some aortic diseases are infrequent, even rare. For instance, tuberculosis or syphilis are rare as the cause of aortic disease in the United States. Aortic aneurysms form when the artery dilates like a balloon. As distinct from aortic dissection, the initiating event is not a tear in the wall of a blood vessel. The clinical course of aneurysmal dilation of the aorta is different from that of aortic dissection and beyond the scope of this course. In general, the urgency of addressing aortic aneurysms surgically depends on the degree of aortic enlargement, rate of change in diameter of the vessel, and the etiology of the disease process. The force of blood pushing against the walls of an artery combined with damage or injury to the artery’s walls can cause an aneurysm.
Many conditions can damage and weaken the walls of the aorta and have been implicated as the cause of aortic dissection. These are just a few of the causes of increased aortic wall stress: uncontrolled hypertension, cocaine or other stimulant use, weight lifting or other Valsalva maneuver, or trauma, such as in a deceleration injury.

Conditions associated with aortic media abnormalities fall into three categories: genetic, inflammatory vasculitides, and other. Examples of genetic conditions include Marfan syndrome, Ehlers-Danlos syndrome (the vascular type), Turner syndrome, and Loeys-Dietz syndrome. These genetic conditions can weaken the body’s connective tissues and can damage the aorta. People who have these conditions may develop aneurysms at a younger age than other people and are at higher risk for rupture and dissection of the aorta.
# Risk Factors for Development of Thoracic Aortic Dissection

## Conditions associated with increased aortic wall stress

- Hypertension, particularly if uncontrolled
- Pheochromocytoma
- Cocaine or other stimulant use
- Weight lifting or other Valsalva maneuver
- Trauma
- Deceleration or torsional injury (eg, motor vehicle crash, fall)
- Coarctation of the aorta

## Conditions associated with aortic media abnormalities

- Genetic
  - Marfan syndrome
  - Ehlers-Danlos syndrome, vascular form
  - Bicuspid aortic valve (including prior AV replacement)
  - Turner syndrome
  - Loeys-Dietz syndrome
  - Familial thoracic aortic aneurysm and dissection syndrome
- Inflammatory vasculitides
  - Takayasu arteritis
  - Giant cell arteritis
  - Behçet arteritis
- Other
  - Pregnancy
  - Polycystic kidney disease
  - Chronic corticosteroid/immunosuppression agent admin.
  - Infections involving the aortic wall either from bacteremia or extension of adjacent infection

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Hiratzka et al 2010 Guidelines on Thoracic Aortic Disease e301
It is recommended that first degree relatives of those affected by aortic dissection undergo genetic screening. Many genetic defects have been identified that are associated with this disease.

Knowledge of whether a first degree relative carries this type of defect will determine if additional testing is necessary and dictate how those affected with this genetic problem should be monitored by their health care providers.
The physical examination should also include a search for the physical stigmata of Marfan syndrome. This is a systemic connective tissue disorder. It involves the cardiovascular, skeletal, and ocular systems.

There are **three cardinal criteria** for the diagnosis of classical Marfan syndrome:

1. **Skeletal features** (long, thin extremities with loose joints, tall and thin in body habitus, arachnodactyly (long slender spider-like fingers), pectus deformities, Scoliosis, a long, thin face, flat feet, risk for spontaneous pneumothorax, and stretch marks)
2. **Dislocation of the lens of the eye** (ectopia lentis) with myopia and an increased risk for retinal detachment
3. **Thoracic aortic disease**
Marfan Syndrome
Skeletal Features

- Long, thin face
- Tall and thin in body habitus
- Long, thin extremities with loose joints
- Arachnodactyly (long, slender, spider-like fingers)

Additional Skeletal Features Not Shown:

• Flat feet
• Scoliosis
• Stretch marks
Cardinal criteria for the diagnosis of classical Marfan syndrome includes dislocation of the lens of the eye (ectopia lentis), with myopia and an increased risk for retinal detachment. This eye diagnosis usually requires pupillary dilation. The ocular globe is usually elongated. These patients are usually myopic.
Marfan Syndrome: Heart

Cardinal criteria for the diagnosis of classical Marfan syndrome includes thoracic aortic disease that begins at the base of the aorta, is located at the aortic heart valve, and extends upward involving the thoracic aorta.

Most Marfan patients die of thoracic aortic dissection if their aortic aneurysm is left undiagnosed and untreated.
Ehlers-Danlos Syndrome, Vascular Form or Type IV

This is a rare autosomal dominant disorder characterized by easy bruising, thin skin with visible veins, characteristic facial features, and a tendency for rupture of arteries, uterus, or intestines. These patients have tissue fragility, a tendency to hemorrhage extensively, and poor wound healing that complicate surgical repair of any type of tissue rupture. Arterial ruptures lead to decreased life expectancy. Median life expectancy for these individuals is 48 years.
Bicuspid Aortic Valve Disease

The aortic valve is a one-way valve between the heart and the aorta, the main artery from the heart that distributes oxygen-rich blood to the body. Usually, the aortic valve has three small flaps or leaflets that open and close to regulate blood flow, allowing blood to flow from the heart to the aorta, and preventing blood from flowing backwards into the heart.

In bicuspid aortic valve disease (BAVD), the valve has only two leaflets. With this abnormality, the valve doesn’t function properly, but it may function adequately for years without causing symptoms or obvious signs of a problem.

The latest studies suggest that bicuspid aortic valve disease is caused by a connective tissue disorder that also causes other circulatory system problems. People with bicuspid aortic valve disease also may have abnormal coronary arteries, aortic aneurysm or an abnormal thoracic aorta, and labile blood pressure. About 2% of the population has BAVD. It is found to be twice as common in males as in females.

The underlying connective tissue disorder that causes BAVD also affects the tissue layers that form the walls of the aorta. As the tissue layers degenerate, the aorta walls lose their strength and stretch out of shape. Eventually, this can cause the formation of an aneurysm, a bulging out area of the aorta where the walls are thin and over-stretched. The aneurysm can rupture or aortic dissection can occur. Rupture and dissection are both life-threatening emergencies.
Turner Syndrome

This syndrome is characterized by complete or partial absence of the female sex chromosome. The most common finding is ovarian failure and short stature. Women with this syndrome have an increased cardiovascular mortality from both structural and ischemic heart disease, especially AoD. Between 10% and 25% of patients with Turner syndrome have a bicuspid aortic valve. The average age of AoD in Turner syndrome is 31 years. Less than half of the patients survived the event.
Features of this syndrome include skeletal features similar to Marfan syndrome, thoracic aortic disease with possible aneurysm and/or rupture. Some patients also have craniofacial abnormalities, cleft palate, ocular hypertelorism, which is an abnormal increased distance between the eyes, and craniosynostosis which is premature closure of the sutures of the skull. These patients can also have thin skin with clearly visible veins and abnormal scars. This diagnosis can be confirmed by molecular genetic testing. Surgical procedures in these patients are not complicated by tissue fragility.
Familial Thoracic Aortic Aneurysm and Dissection Syndrome

There is a strong genetic component in thoracic aortic dissection as depicted in the Tyler Kahle video.

Dianna M. Milewicz, MD, PhD, is the Vice Chair the Department of Internal Medicine and Director of the Division of Medical Genetics, at the University of Texas Medical School at Houston. Dr. Milewicz is currently the President George H. W. Bush Chair of Cardiovascular Medicine. Her research team studied the family of Tyler Kahle and other families similarly affected with aortic dissection. Her research has allowed her to identify the genetic defect in many of these families associated with aortic aneurysms leading to aortic dissections and how the genetic defect is passed on from generation to generation in an autosomal dominant manner.
A genetic predisposition to thoracic aortic disease can be inherited in families associated with syndromic features, as in Marfan syndrome and Loeys-Dietz syndrome. Additionally, up to 20% of thoracic aortic disease patients have a predisposition for thoracic aortic disease inherited in the family without any syndromic features. Both syndromic and non-syndromic thoracic aortic diseases are typically inherited in an autosomal dominant manner and many causative genes have been identified.

It is recommended that first degree relatives of those affected by aortic dissection undergo genetic screening. Many genetic defects have been identified that are associated with this disease. Knowledge of whether a first degree relative carries this type of defect will determine if additional testing is necessary and dictate how those affected with this genetic problem should be followed by their health care providers.
Genetic testing may be completed at larger general hospitals and medical academic institutions. This testing is indicated in thoracic aortic disease patients with syndromic features, a family history of the disease or a young age of onset of aortic aneurysms or dissections. The majority of individuals diagnosed with familial thoracic aortic disease have an affected or carrier parent. The children of an affected parent are at 50% risk of inheriting the mutant gene and the disease.

Once a genetic defect is identified, other family members can be screened for the disease. If they have the defective gene, routine imaging of the aorta is recommended and surgical repair done to prevent an acute aortic dissection. If the causative gene is not identified, first degree family members should be imaged for aortic disease.
Inflammatory Diseases Associated with Aortic Dissection

**Takayasu's Arteritis**
- Known as the pulseless disease because the primary symptom is an absent radial pulse
- Autoimmune inflammatory disease that causes a panarteritis, most often related to thickening and occlusion of larger arteries
- Frequently affects the aortic arch causing an aortic arch dissection or rupture
- Sometimes, this disease involves the ascending aorta
- Found mainly in young Asian women.

**Giant cell Arteritis**
- Known as temporal arteritis, includes a vascular disease which primarily affects women over 50
- Involves medium- and large-sized arteries
- May result in obstruction and inflammation of the temporal and ophthalmic arteries
- Giant cell arteritis might involve the aorta, with the development of an inflammatory aortitis
- This usually does not progress to aneurysm formation
- The pathology includes the focal granulomatosis lesions, involving the entire arterial wall
- There is a genetic predisposition to this disease
- Patients complain of headaches and malaise
- They also have an elevated sedimentation rate and anemia
- The thickened nodular temporal artery, anterior to the ear, may feel tender to touch

**Beçhet Arteritis**
- Characterized by genital ulceration, uveitis or retinal vasculitis, or skin lesions
- Patient may also manifest vascular abnormalities
- This vasculitis may affect any veins or arteries, big or small
- Though unusual for patients Beçhet Arteritis to have aortic involvement, aortic histopathology has shown lymphocytic infiltration mixed with histiocytes and eosinophils with giant cells around vasa vasorum of media and adventitia. Destruction of the media can lead to aneurysm formation and may proceed to pseudoaneurysm formation and rupture.
Other Causes of Aortic Dissection

- Pregnancy
- Polycystic kidney disease
- Chronic corticosteroid or immunosuppression agent administration
- Infections involving aortic wall or extension of adjacent infection
- Trauma
Pregnancy

- Marfan syndrome, Ehlers-Danlos syndrome, and other non-Marfan aortic disease may manifest during pregnancy.
- Although clearly rare, it has been estimated that half of AoD and/or ruptures in women younger than 40 years of age have been associated with pregnancy.
- Most dissections occur in the ascending aorta, although dissection or rupture of virtually any artery in the body has been described.
- Additionally, pregnancy-related expansion of the sinotubular junction may lead to aortic valve insufficiency.
- Complications include rapid progression of aortic root or ascending aortic enlargement or aortic dissection without prior enlargement of the aorta.
- Aortic dissection or rupture may occur during pregnancy, delivery, or even post-partum.
- Women with genetic connective tissue diseases, known thoracic aortic disease or a family history of thoracic aortic aneurysms or dissections should be considered for pre-pregnancy counseling from cardiologists, genetics, and high-risk obstetricians.
Recommendations for Counseling and Management of Chronic Aortic Diseases in Pregnancy

Class I

• Women with Marfan syndrome and aortic dilatation, as well as patients without Marfan syndrome who have known aortic disease, should be counseled about the risk of aortic dissection as well as the heritable nature of the disease prior to pregnancy. (Level of Evidence: C)

• For pregnant women with known thoracic aortic dilatation or a familial or genetic predisposition for aortic dissection, strict blood pressure control, specifically to prevent Stage II hypertension, is recommended. (Level of Evidence: C)

• For all pregnant women with known aortic root or ascending aortic dilatation, monthly or bimonthly echocardiographic measurements of the ascending aortic dimensions are recommended to detect aortic expansion until birth. (Level of Evidence: C)

• For imaging of pregnant women with aortic arch, descending, or abdominal aortic dilatation, magnetic resonance imaging (without gadolinium) is recommended over computed tomographic imaging to avoid exposing both the mother and fetus to ionizing radiation. Transesophageal echocardiogram is an option for imaging of the thoracic aorta. (Level of Evidence: C)

• Pregnant women with aortic aneurysms should be delivered where cardiothoracic surgery is available. (Level of Evidence: C)

Class IIa

• Fetal delivery via cesarean section is reasonable for patients with significant aortic enlargement, dissection, or severe aortic valve regurgitation. (Level of Evidence: C)

Class IIB

• If progressive aortic dilatation and/or advancing aortic valve regurgitation are documented, prophylactic surgery may be considered. (Level of Evidence: C)
Polycystic kidney disease (PKD) is a genetic disorder characterized by the growth of numerous cysts in the kidneys. The kidneys are two organs, each about the size of a fist, located in the upper part of a person's abdomen, toward the back. The kidneys filter waste and extra fluid from the blood to form urine. They also regulate amounts of certain vital substances in the body. When cysts form in the kidneys, they are filled with fluid. PKD cysts can profoundly enlarge the kidneys, replacing much of the normal structure, resulting in reduced kidney function and subsequently leading to kidney failure.

PKD can also cause cysts in the liver and problems in other organs, such as blood vessels in the brain and heart. The number of cysts as well as the complications they cause help doctors distinguish PKD from the usually harmless "simple" cysts that often form in the kidneys in later years of life.
People with autosomal dominant PKD can also experience the following complications:

- Urinary tract infections - specifically, in the kidney cysts
- Hematuria (blood in the urine)
- Liver and pancreatic cysts
- Abnormal heart valves
- High blood pressure
- Kidney stones
- Aneurysms (bulges in the walls of blood vessels) - in the brain
- Diverticulosis (small pouches that bulge outward through the colon)
There have been untold benefits of corticosteroids and immunosuppressive agents in the treatment of myriad diseases. Patients who take these medication on an ongoing basis may suffer high incidence of premature atherosclerosis. There is also evidence of increased rates of vascular mortality in corticosteroid-treated diseases, such as rheumatoid arthritis, reversible airway obstruction and transplant recipients. Possible mechanisms of damage include elevated blood pressure, impaired glucose tolerance, dyslipidemia, and imbalances of thrombosis and fibrinolysis.
Infections Involving Aortic Wall or Extension of Adjacent Infection

- These infections develop from
  - rare fungi,
  - staphylococcal,
  - streptococcal, or
  - salmonella aortic infections
- Usually originates as an atherosclerotic plaque of an aneurysm
  - These are saccular type aneurysms
- The blood cultures will be positive for bacteria and/or fungi
- In third world countries, TB and syphilis would also be considerations
Trauma

- Traumatic aneurysms occur after penetration or acceleration/deceleration injuries.
- These traumatic aneurysms commonly affect the descending thoracic aorta.
- Blunt force trauma with rapid deceleration type injuries, such as car accidents or falls, can also damage the walls of the aorta and lead to TAAs.
- The descending thoracic aorta is highly susceptible to these types of injuries.
- The mechanism of injury is associated with a combination of shearing forces, compression of the aorta on the vertebral column or manubrium, and an increase of pressure inside the vessel during the episode of the trauma.
- The majority of patients with blunt aortic injuries who survive to reach the hospital sustain an injury in the isthmus of the aorta between the left subclavian artery and the ligamentum arteriosum.
Module 3 Review Questions

Before moving on to Module 4, please answer the review questions for Module 3.
Module 4: Patient Presentation
Presenting Symptoms of Aortic Dissection

Pain

- **Abrupt onset** of pain
- Ascending aortic dissection typically produces substernal pain
- Descending dissection produces posterior back pain in the intrascapular region
- Patients descriptively report the pain as “splitting” or “tearing” in nature
- Other symptoms may reflect ischemia of the particular vascular branches that may be impaired or occluded by the progressing dissection
- Pain is usually the most intense it will ever be at onset, then,-curiously, it can lessen or even go away, creating a FALSE sense of security about the severity of the condition

Capability of Producing Ischemia of the Brain

- Resulting in
  - Stroke
  - Ischemia of the spinal cord, resulting in paraplegia
  - Ischemia of the intestines, resulting in bowel necrosis;
  - Ischemia of the kidneys, resulting in renal failure or renovascular hypertension
  - Iliac artery ischemia, resulting in pulseless lower extremities
Four Means by Which Aortic Dissection Can Lead to Death

- Intrapericardial rupture, producing cardiac tamponade
- Free intrapleural rupture, usually on the left side
- Acute aortic insufficiency, resulting in acute cardiogenic shock
- Vascular compromise, from any of the indicated branch arteries of the aorta
Sir William Osler, considered to be the father of modern medicine, emphasized the importance of listening to the patient, saying, “If you listen carefully to the patient they will tell you the diagnosis.” In the case of aortic dissection, however, one might well add the stipulation, “provided that you ask the patient the right questions.”

Often, the patient is asymptomatic in the early stages of thoracic aortic disease. Histories and symptoms can vary greatly from one patient to another. This difference can exist even with identical pathology and stage of the disease adding to the difficulty in making the proper diagnosis. No one sign or symptom can identify an acute dissection.

Patients may present with complaints of chest pain ranging from mild anterior pain to tearing, ripping pain or no even pain at all. They may also present with vaguer symptoms such as nausea, fever, altered mental status, syncope or near syncope, dyspnea, hemoptysis, flank pain, abdominal pain, parathesias in the limbs or a weakness on one side.

Stroke symptoms may also be the presenting complaint. Patients may have a combination of several symptoms or just a single symptom. A good strategy for early diagnosis, and possibly patient survival, is to assume all patients presenting with these type symptoms have a thoracic aortic dissection until proven otherwise. **In-depth histories and physical assessments are key.**
There is a misconception in the medical community that aortic dissection is a disease of the older adult only. This is not the case. Aortic dissection can occur in patients of any age. Even toddlers have been reported with aortic dissection, Piskac (2008).

In the acute dissection stage of thoracic aortic disease, the classic patient presentation is sudden onset of severe chest pain. This type of thoracic pain has been described as tearing, sharp, stabbing or ripping in presentation. The pain may be retrosternal, radiating to the neck or throat, or localized to the anterior or posterior chest areas. Some male patients may also complain of testicular pain. The patient may present with a cough, sob, hoarseness, or dysphasia. The pain may also subsequently ease or abate, leading to a false reassurance on the part of the patients and the physicians. The pain may also migrate or change as the tear or dissection progresses. Other associated symptoms may include syncope, dizziness, malaise, general weakness, or even unconsciousness. There may be additional history and complaints referable to compression of adjacent tissues, for example, flank pain from compression of the renal artery. Lastly in the emergency room scenario, the patient may be hypotensive, or unresponsive requiring immediate surgery.

Patients may not easily volunteer a history that significantly increases the probability that aortic dissection is the etiology of their chest pain. Hence, the ideal evaluation of chest pain in the emergency room should include questioning of the patient regarding a family history of aortic dissection or a personal history of Marfan syndrome or other connective tissue disorders. The provider must also maintain a high suspicion for thoracic aortic dissection until proven otherwise.
Various Possible Physical Findings

A dilated thoracic aorta may compress other tissues which may lead to both symptoms and physical findings.

- Chest Pain
- Dyspnea
- Cough
- Hoarseness
- Dysphasia

Acute dilatation of the thoracic aorta causes aortic valve regurgitation resulting from aortic valve diameter increases and therefore incompetence.

- Peripheral pulmonary rales
- Peripheral bounding pulses
- A wide pulse pressure
- A diastolic murmur along the right sternal border

Dilation and dissection of the proximal thoracic aortic artery:

- Compression/ constriction of the coronary artery orifices
- Decreased coronary blood flow
- Myocardial ischemia and infarct

A dissection or dilatation of the thoracic aortic aneurysm and thoracic aortic arch:

- May compress & constrict the very proximal segments of the right innominate, the left common carotid, and the left subclavian arteries

Possible results of aortic valve regurgitation:

- Compromise of the carotid circulation could have serious neurological sequelae such as ischemia, infarct, and/or stroke
Aortic dissection is missed in up to 38% of patients on initial evaluation, and in up to 28% of patients the diagnosis is made at autopsy.\textsuperscript{2,4} Traditionally, aortic dissection without pain was thought to be rare. More recent information suggests that symptoms in patients with aortic dissection are more variable than previously recognized, and the classic findings of sudden onset of tearing chest, back, or abdominal pain are often absent.
A dilated expanding thoracic aorta might cause superior vena cava syndrome occurring secondary to superior vena cava area obstruction. The pathophysiology of the superior vena cava obstruction occurs from the obstruction of the venous return from the head, neck, and upper extremities. The physical findings of the superior vena cava syndrome include face and neck swelling with discoloration of a bluish-purple color, dyspnea, coughing, tongue edema, hoarseness, nasal congestion, nasal bleeding, and hemoptysis. Dilated facial and neck veins are prominent. These physical manifestations are accentuated with the patient in a supine position.

If the superior cervical ganglia is compressed by the dilated aorta, there would also be the findings of Horner's syndrome. Horner’s syndrome results from an interruption of the sympathetic nerve supply to the eye and is characterized by the classic triad of miosis (i.e. constricted pupil), partial ptosis (an abnormal low-lying upper eyelid margin with the eye in primary gaze), and anhidrosis (loss of hemi-facial sweating).
Rapid bedside screening for acute aortic dissection can be easily accomplished by application of the Acute Dissection Detection Score (ADD Score).

This score was derived from registry data that demonstrate that the vast majority of aortic dissection occurs in patients with easily identified risk features.

The score can be used at the bedside and requires no laboratory or imaging studies.

It has been shown to be 96% sensitive for identification of patients with aortic dissection. *

Source: Dr. Luke Hermann 8-29-2012

* Circulation 2011, 123:2213-2218
The risk features that comprise the score can be broken down into three simple categories as follows:

- **C**: Conditions that predispose to aortic dissection
- **P**: Pain features that predict aortic dissection
- **E**: Exam features consistent with aortic dissection

Source: Dr. Luke Hermann 8-29-2012
Conditions that predispose to aortic dissection

**Congenital Syndrome**
- Marfan Syndrome
- Ehlers Danlos Syndrome
- Loeys Deitz Syndrome
- Turner Syndrome

**Aortic Conditions**
- Bicuspid valve
- Family history dissection
- Instrumentation*
- Thoracic aneurysm

* Recent surgery involving aorta or cardiac catheterization.

Source: Dr. Luke Hermann 8-29-2012
pain features that predict aortic dissection

- pain that is **abrupt** in onset
- pain that is **severe** in intensity
- pain described as **ripping** or **tearing**
Exam features consistent with aortic dissection

- perfusion deficit
- neurologic (stroke)
- mesenteric (abdominal pain)
- myocardial (heart attack)
- peripheral (ischemic limb)
- murmur of aortic insufficiency

Source: Dr. Luke Hermann 8-29-2012
taken together, these 3 categories make up the ADD Risk Score

The ADD Risk Score is calculated as follows:

C: conditions that predispose to aortic dissection
   The presence of one or more high risk conditions = 1 point.

P: pain features that predict aortic dissection
   The presence of one or more high risk pain features = 1 point.

E: exam features consistent with aortic dissection
   The presence of one or more high risk exam features = 1 point.
Once calculated, the risk score can then be used to guide further testing.

- **Patient High Risk**
  - 2 or more points
  - Immediate Imaging Study

- **Patient Moderate Risk**
  - 1 Point
  - Immediate Imaging Study if initial evaluation not diagnostic of alternate cause of symptoms

- **Patient Low Risk**
  - 0 Points
  - Consider imaging study if initial evaluation not diagnostic of alternate cause of symptoms

Source: Dr. Luke Hermann 8-29-2012
The evaluation of patients with symptoms that could represent aortic dissection is summarized in the following adapted graphic which was created for 2010 American Heart Association / American College of Cardiology Guideline on Thoracic Aortic Disease. *

* Circulation. 2010; 121: e266-e369
Consider acute AoD in all patients presenting with:
- Chest back, or abdominal pain
- Syncope
- Symptoms consistent with perfusion deficit (i.e. CNS, mesenteric, myocardial, or limb ischemia)

**STEP 2**
Bedside risk assessment

**Focused bedside pre-test risk assessment for acute AoD.**

1. **High Risk Conditions**
   - Marfan Syndrome
   - Connective tissue disease
   - Family history aortic disease
   - Known aortic valve disease
   - Recent aortic manipulation
   - Known thoracic aortic aneurysm

2. **High Risk Pain Features**
   - Chest, back, or abdominal pain described as the following:
     - Abrupt in onset/ severe in intensity
     - Rippling/tearing/sharp or stabbing quality

3. **High Risk Exam Features**
   - Evidence of perfusion deficit
   - Pulse deficit
   - Systolic BP differential
   - Focal neurologic deficit (in conjunction with pain)
   - Murmur of aortic insufficiency (new or not known to be old and in conjunction with pain)
   - Hypotension or shock state

Determine pre-test risk by combination of risk conditions, history, and exam.

**STEP 3**
Risk based diagnostic evaluation

- **Low Risk**
  - No high risk features present.

- **Intermediate Risk**
  - Any single high risk feature present.

- **High Risk**
  - Two or more high risk features present.

Hiratzka et al 2010 Guidelines on Thoracic Aortic Disease e310
Determine pre-test risk by combination of risk conditions, history, and exam.

**STEP 3**
Risk based diagnostic evaluation

- **Low Risk**
  - No high risk features present.
  - Proceed with diagnostic evaluation as clinically indicated by presentation.
  - Alternative diagnosis identified?
    - Yes: Initiate appropriate therapy.
    - No: Continue with diagnostic evaluation.

- **Intermediate Risk**
  - Any single high risk feature present.
  - EKG consistent with STEMI?
    - Yes: Likely primary ACS. In absence of other perfusion deficits, strongly consider immediate coronary re-perfusion therapy. If coronary angiography performed is culprit lesion identified?
      - Yes: Initiate appropriate therapy.
      - No: Continue with diagnostic evaluation.
    - No: CXR with clear alternate diagnosis?
      - Yes: Initiate appropriate therapy.
      - No: History and physical exam strongly suggestive of specific alternate diagnosis?
        - Yes: Alternate diagnosis confirmed by further testing?
          - Yes: Initiate appropriate therapy.
          - No: Continue with diagnostic evaluation.
        - No: Consider aortic imaging study for TAD based on clinical scenario (particularly in patients with advanced age, risk factors for aortic disease, or syncope).

- **High Risk**
  - Two or more high risk features present.
  - Immediate surgical consultation and arrange for expedited aortic imaging.

**STEP 4**
Acute AoD identified or excluded

- If high clinical suspicion for aortic dissection exists, consider secondary imaging study.
- Aortic Dissection Present?
  - Yes: Proceed to Treatment Pathway
  - No: Aortic Imaging Study
    - TEE (preferred if clinically unstable)
    - CT
    - MR chest to pelvis

Hiratzka et al 2010 Guidelines on Thoracic Aortic Disease e310
The management of patients with symptoms that could represent aortic dissection is summarized in the following graphic which was created for 2010 American Heart Association / American College of Cardiology Guideline on Thoracic Aortic Disease. *

* Circulation. 2010; 121: e266-e369
Hiratzka et al 2010 Guidelines on Thoracic Aortic Disease e311

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*Continued from previous page
Radiology and Laboratory Testing

A normal PA and lateral chest radiograph does not rule out the presence of aortic dissection. Hence, the chest X-ray alone is inadequate as an imaging technique to evaluate patients at high risk for aortic dissection. A widened mediastinum and a widened aortic silhouette are classic findings. Left-sided pleural effusions may also be present. Hemothorax may also be noted if the dissection has ruptured.

The gold standard for detection of aortic dissection is computed angiographic tomography of the chest (CT angiography). This modality is widely available, can be rapidly obtained, is expedient and accurate. Other acceptable imaging modalities include magnetic resonance angiography (MRI angiography) and transesophageal echocardiogram (TEE).

If a single imaging study is not conclusive in a high risk patient defined from the evaluation table, strong consideration should be given to obtaining a second imaging study. The mortality rate for acute aortic dissection increases by 2% for every hour delay in making the diagnosis as cited by Geirsson, et.al. (2007). Hence, emergent imaging by an acceptable modality is indicated in patients at high risk for aortic dissection with compatible symptoms. Survival may depend on rapid diagnosis and surgical treatment.
Dissection of the ascending aorta is a surgical emergency.

Initial management of thoracic aortic dissection should be directed at maintaining A, B, C’s and decreasing aortic wall stress by controlling heart rate and blood pressure as follows:

A. In the absence of contraindications, intravenous beta blocker should be initiated and titrated to maintain a target heart rate of 60 bpm or less. Commonly used medications are Esmolol and Labetolol. In patients with a clear beta blockade contraindication, a nondihydropyridine calcium channel blocker, such as Diltiazem or Verapamil should be used as an alternative for rate control.

B. If systolic BP remains greater than 120 mm Hg after adequate heart rate control has been obtained, then ACE inhibitor and/or other vasodilators should be administered intravenously to further reduce blood pressure that maintains adequate end-organ perfusion. A common medication used is Nipride.

C. Beta blockers should be used cautiously in the setting of acute aortic regurgitation because they will block the compensatory tachycardia.

D. Vasodilator therapy should not be initiated prior to rate control so as to avoid associated reflex tachycardia that may increase aortic wall stress, leading to the expansion of a thoracic aortic dissection.
Pay particular attention to the patient’s history, especially the PQRST of the patient’s pain as this may change with increases in tears/dissections. Place the patient in Semi-Fowler’s position for comfort. (You want to alleviate as much anxiety as possible and provide comfort).

When assessing a patient who complains of chest pain that may be an aortic dissection, if the systolic blood pressure is significantly different between the right and left arm, the carotid, radial, and femoral pulses should be palpated for any abnormal findings. If these pulses are diminished or absent, this sign may indicate an aortic dissection (Dixon, 2011). In the patient who has a descending aortic dissection, peripheral pulse deficits would be distal to the dissection according to McCallren-Currey, Shaughnessey, K. (1999).
Nursing Physical Assessment Pearls

**Inspection**

1. Assess the patient’s level of consciousness, behavior, affect, or are they comatose?

2. Compare the blood pressures in not only the bilateral upper extremities, but also the lower extremities. Blood pressures will be significantly greater in the upper extremities than the lower. Any 20mmHg difference between the right upper and left upper extremity should be investigated further.

3. Is hypertension present? Hypotension? (Hypertension may be seen with distal dissections and hypotension is common with proximal dissections).

4. Is tachycardia present?

5. Is the patient in severe distress or anxious?

6. Is the patient diaphoretic?

7. Is pallor present?

8. Peripheral cyanosis present?
Nursing Physical Assessment Pearls

**Palpation**

- Are pulse deficits/decreased peripheral pulses noted?
- Are the upper extremity pulses bounding? (Found in acute aortic regurgitation)
- Pulsatile masses noted?
- Are there any parathesias/hemiplegias/paraplegias present? (Continually reassess as it can change with increasing tears/dissections.)
- Diffuse tenderness noted?
Nursing Physical Assessment Pearls

Auscultation

Is there a murmur noted to the right of the sternal border?

Breath sounds with rales?

Muffled heart tones?

(cardiac tamponade)
Nursing Physical Assessment Pearls (Continued)

Nursing initial priorities are to maintain the A,B,C’s (airway, breathing, and circulation)

- Provide supplemental oxygen to the patient
- Consider the possible need for proactive intubation
- Place two large caliber IV or IO catheters
- Critically assess the probability of dissection and the need for giving volume/packed red blood cells (PRBCs)
- Initiate cardiac and pulse oximetry monitoring
- Prepare for immediate surgery with indwelling foley catheter and NG tube placed to low intermittent wall suction
- Keep the patient calm, speak gently but informatively
- Give pain medications and other pharmacologic meds as ordered

When the diagnosis of aortic dissection has been confirmed by an appropriate imaging modality, an immediate consultation with a cardiothoracic surgeon is indicated, regardless of the anatomic location. Keep in mind, for every hour delay in surgery, the mortality rate of Class A thoracic aortic aneurysm dissection increases by 2%.
Definitive treatment of Type A dissections is surgical repair. Once the patient is taken to surgery and the chest cavity is opened, the patient is placed on a heart-lung machine and cooled to help preserve brain and other vital organ function. A graft is then employed to replace the section of the aorta where the tear occurred. The circulation may be stopped for a period of time to facilitate the repair, particularly of aortic arch arteries.

The dissection may extend into the very proximal ascending aorta, involve the origin of the coronary arteries, or result in incompetence of the aortic valve. Surgical repair in such instances may require that the coronary origins be reattached to the aortic graft or that the aortic valve be replaced.

Additional complicating features arise when the great vessels of the aortic arch are involved with the dissection. Grafts to the innominate, carotid, and subclavian vessels may be required, procedures that extend the time of circulatory arrest.
Type B dissections are usually not treated surgically. Possible indications for surgery in Type B dissections include vessel leaking, rupture, or compromise of blood flow to vital organs. Those with Marfan syndrome are usually treated more aggressively with surgery.

The surgical repair of Type B dissection more often results in paraplegia than does that of Type A dissection due to disruption of the segmental blood supply to the spinal cord.

Medical treatment of Type B dissections consists of controlling high blood pressure and utilization of beta blocker medications that decrease the rate of change of pressure in the aorta over time. These measures minimize stress to the affected vessels.
Aortic dissection can occur at any age. The commonly held notion that aortic dissection is a disease solely of the elderly or adult is incorrect. Acute dissection of the ascending aorta (Type A dissection) is a surgical emergency. Each hour that a Type A aortic dissection is delayed, the mortality rate increases by 2%.

Aortic dissection occurs more frequently than is believed by both the medical community and the lay public. As many as 15,000 people die each year in the United States from this illness. It is believed by experts that in many, if not most of these deaths, the etiology is not recognized before death occurs. The key to decrease mortality is to diagnose and treat early.

Early recognition, and ultimately diagnosis, is accomplished by maintaining a high suspicion for high risk patients. High-risk patients include those with a family history of aortic dissection or aneurysms, patients diagnosed with Marfan syndrome, and those with physical characteristics of Marfan syndrome or other connective tissue disorders (having “syndromic features”), and patients with a bicuspid aortic valve, including previous aortic valve replacement.

Utilizing the most appropriate diagnostic tool for either “ruling in” or “ruling out” thoracic aortic dissections in high risk patients is a necessity. CT angiogram (the gold standard), MRI angiogram, or Transesophageal echocardiography are the most effective diagnostic tools. It should be noted in high risk patients that if the first diagnostic study is negative, a second imaging study is warranted. Although chest X-rays are necessary, they are not definitive diagnostics, as a negative x-ray may still be positive for aortic dissection.
Medical staff can often miss the diagnosis of thoracic aortic dissection because both the symptoms of thoracic aortic dissection and the ages of presenting patients can vary greatly. The classic symptoms of aortic dissection (acute sharp or tearing chest pain) may not be present in high-risk patients. Proper imaging of the chest is the only means currently available to exclude aortic dissection in high-risk patients with symptoms compatible with aortic dissection.

Patients often do not volunteer a family history of aortic dissection, Marfan syndrome, or other high risk conditions and history. To identify those at high risk for aortic dissection, the triage/intake evaluation of patients with chest pain ideally should require that the providers actively inquire about a personal history or family history of these conditions. High risk patients include those with a family history of aortic dissection, Marfan syndrome or other connective tissue disorders, or syndromic features of these disorders. Because risk of the disease may be passed from parent to offspring, first-degree relatives of patients with aortic dissection should be considered for genetic testing for conditions that may predispose them to aortic aneurysm or dissections.
Dr. Milewicz's and others work in identifying a molecular biomarker to identify acute dissections for use in the emergency scenario is ongoing. The ability to provide a rapid diagnosis and surgical intervention will increase survival rates.

Nebraska Methodist Hospital is committed to promoting a nationwide change in chest pain protocols

Methodist Health System, together with our fellow TAD Coalition members, are working for nationwide implementation of the 2010 AHA/ACC diagnostic and treatment guidelines.

Please review and implement these clinical guidelines, which are more comprehensive. The full 108-page text and free copies of the pocket version of the aortic dissection clinical guidelines are available through the TAD Coalition at www.tadcoalition.org.
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