An Aortic Dissection: The Adam Carabajal Story
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Adam’s Story
- History
- July 19, 2009
- Signs and symptoms
- EKG, Labs, X-rays
The Aorta

- The aorta is the largest artery in the body and is the blood vessel that carries oxygen-rich blood away from the heart to all parts of the body.
- The section of the aorta that runs through the chest is called the thoracic aorta and, as the aorta moves down through the abdomen, it is called the abdominal aorta.
- Can be divided into several sections:
  - Aortic root
  - Ascending aorta
  - Aortic arch
  - Descending aorta
  - Abdominal aorta

Thoracic Aortic Aneurysms

- When an artery wall in the aorta weakens, the wall abnormally expands or bulges as blood is pumped through it, causing an aortic aneurysm.
- When this defect develops, blood pressure can force the bulge to tear open and allow blood to pass through.
- Since the blood is under pressure, it eventually splits (dissecting) the middle layer of the blood vessel, creating a new channel for blood. The length of the channel grows over time and can result in the closing off of connection points to other arteries.
- This can lead to heart attack, strokes, abdominal pain, and nerve damage. Blood may leak from the dissection and collect in the chest or around the heart.
Thoracic Aortic Aneurysms

- Aneurysms that occur in the section of the aorta that runs through the abdomen (abdominal aorta) are called abdominal aortic aneurysms.
- Aortic aneurysms that occur in the chest area are called thoracic aortic aneurysms and can involve the aortic root, ascending aorta, aortic arch or descending aorta. Aneurysms that involve the aorta as it flows thru both the abdomen and chest are called thoracoabdominal aortic aneurysms.

Signs and Symptoms

- Sudden, intense pain
- Pain so intense as to immobilize the patient and cause him to fall to the ground
- The pain in both the chest and in the back, between the shoulder blades
- Dyspnea
- Cool and Clammy
- Diaphoretic
- Burning sensation in upper chest
- Hypotension
- Hypertension
- Tachycardia
- Nausea and vomiting

- Swelling in neck or arms
- Aortic aneurysms usually do not cause any symptoms until they are quite large.
- Coughing and wheezing
- Hoarse voice
- Difficulty swallowing
- Severe onset of sharp, stabbing, tearing or ripping pain usually in the chest (front, back or both) or lower back depending on the site of the dissection.

- Horner's syndrome (constricted pupil, drooping eyelid and dry skin on one side of the face)
- Deep boring pain or pulsation in the lower back region
- Decreased ability to move or decreased sensation in an arm or leg or other part of the body.
- Fainting shortness of breath, weak or absent pulse.
- In may be no pain but a sense that there is something terribly wrong.
TAA quick facts

• Usually asymptomatic and not easily detectable
• When detectable usually catastrophic complication occurs
• CT scans, MRI, Echocardiograms, Abdominal ultrasound (to look for associated abdominal aneurysms)
• Angiography (an x-ray of the blood vessels)
• Mimics heart attack
• Can be misdiagnosed for Pleuraly
• Age is never a factor

Causes

• Trauma to chest wall
• Marfans Syndrome
• Ehlers-Danlos syndrome
• Turner Syndrome
• Loey-Dietz syndrome (LDS)
• Bicuspid Valve disease
• Familial Aortic Aneurysms
• A history of personal or family history of thoracic disease
• Rarely Pregnancy
• Connective tissue disorders
### Types of Dissections

- Dissections are also categorized by their location on the aorta where they occur.
- Type A or proximal dissections begin at the ascending aorta.
- Type B or distal dissections begin in the descending aorta and can extend down into the abdominal aorta.
- Other categories used to describe location on the aorta are type I, II and III.
- Type I originates in the ascending aorta, continues to the aortic arch and often beyond it.
- Type III: originates in the descending aorta and extends down the aorta or, rarely, up into the aortic arch and ascending aorta.

### Marfan’s Syndrome

- Disorder of the connective tissue.
- Found in the heart, blood vessels, bones, joints, and eyes.
- Caused by a defect in gene that tells the body how to make fibrillin – a protein that is an important part of connective tissue.
- Increased in a protein called transforming growth factor beta.
- May get worse as people age.

#### Heart and Blood

- Enlarged or bulging aorta
- Separation of the layers of the aorta that can cause it to tear (aortic dissection)
- "Hippo" mitral valve

### Ehlers-Danlos syndrome

- Connective tissue disorders characterized by articular hypermobility, skin extensibility and tissue fragility.
- EDS are due to faulty collagen. Collagen is a protein that acts like glue in the body adding strength and elasticity.
- There are six major types of EDS.
- Vascular (formerly EDS Type IV) is generally regarded as the most serious form of EDS due to arterial or organ rupture.
- Skin is usually thin and translucent with wrinkles.
- Facial characteristics present in some.
- Include large eyes, thin nose, lobe-less ears, short stature and thin scalp hair.
- Spontaneous arterial rupture has a peak incidence in the third or fourth decade of life.
- Skin and muscle rupture can occur.
- Clubfoot is frequently seen at birth.
Turner Syndrome

- 1 out of every 5,000
- Major heart defects to minor cosmetic issues. Some may have only a few features
- Turner syndrome loss of ovarian function, severity of these problems varies

General Appearance:
- Short neck with a webbed appearance
- a low hairline at the back of the neck, and low-set ears.
- Hands and feet of affected individuals may be swollen or puffy at birth, and often have soft nails that turn upward at the ends when they are older.
- Due to obstruction of the lymphatic system during fetal development.
- Cosmetic feature is the presence of multiple pigmented nevi, which are colored spots on the skin, or moles.

Short Stature:
- due to the loss of one copy of the SHOX gene on the X chromosome. The loss of SHOX may also explain some of the skeletal features
  - short fingers and toes, and irregular rotations of the wrist and elbow joints.
  - Linear growth is long and narrow in size, and statural growth lags during childhood, resulting in adult heights of approximately 4 feet 8 inches.
- Final adult height in Turner syndrome can be increased by several inches if growth hormone (GH) is given relatively early in childhood.

Turner Syndrome Cont.....

Cardiovascular:
- From 5% to 10% of children found to have a severe constriction of the major blood vessel coming out from the heart, a condition known as "coarctation of the aorta."
- This can be surgically corrected as soon as it is diagnosed.
- 30% of individuals with Turner syndrome are reported to have "bicuspid aortic valves," meaning that the major blood vessel from the heart has only two rather than three components to the valve regulating blood flow.
- It requires careful medical monitoring, since bicuspid aortic valves can deteriorate or become infected.

Other components affected by Turner Syndrome:
- Kidney
- Osteoporosis
- Diabetes
- Thyroid
- Cognitive Function/Education issues
- Puberty/Reproduction
Loeys-Dietz syndrome (LDS)

- Connective tissue disorder
- Aneurysms: Enlargement is most often observed in the aortic root but can be seen in other arteries throughout the body
- Twisting or spiraled arteries
- Widely spaced eyes
- Craniofacial:
  - Mandibular hypoplasia
  - Hypertelorism
  - Anteriorly notched chin
- Cardiovascular:
  - Aneurysms
  - Patent ductus arteriosus
  - Hypertrophic cardiomyopathy
- Skeletal:
  - Clubfoot
  - Scoliosis
- Joint:
  - Cervical spine instability
  - Pelvic laxity
  - Narrowing of the thoracic inlet
  - Cleft palate
- Skin:
  - Translucent
  - Soft or velvety skin
  - Easy bruising
  - Abnormal or wide scarring
  - Soft skin texture
  - Hernias

Loeys-Dietz syndrome (LDS) cont....

LDS exhibits an autosomal dominant inheritance pattern. Individual has been diagnosed with LDS, such that their offspring have a 50% chance of inheriting the gene mutation.
There is no way to predict the severity of aortic root aneurysms that may occur in an offspring.
Many individuals are born in the family to have the mutation causing LDS. These cases are caused by sporadic (random) mutations that occur during conception.
Vascular surgery is a widely recommended intervention and is a preventative surgery for individuals with a rapidly enlarging aorta or a pronounced family history of aortic dissection. Aortic root replacement is the most common vascular surgery performed in individuals with LDS. and is highly successful.

Confidentiality Management
- Pressure on the aorta is typically controlled by medications such as beta-blockers.
- Work to lessen the strain on the body's major arteries.

Exercise restrictions are typically put in place to assist in slowing the rate of aortic and arterial aneurysm growth. It is advised that individuals with LDS avoid competitive sports, especially contact sports and other exercises that may produce fatigue.

Bicuspid Aortic Valve Disease

- A bicuspid aortic valve is an aortic valve with only two leaflets instead of three. Current clinical and scientific studies are revealing that bicuspid aortic disease is not a simple congenital abnormality.
- It can affect multiple family members. Some affected family members have bicuspid aortic valves, and some have normal-appearing trifoliate aortic valves.
- Regardless of the number of aortic valve leaflets, all family members should be checked periodically to avoid serious, potentially life-threatening complications of the aorta and vessels of the head and neck.
Familial Aortic Aneurysms

- Familial aortic aneurysm (FAA) refers to a genetic predisposition to thoracic aortic aneurysm or dissection in the absence of other syndromes such as Marfan or Loeys-Dietz. Studies show that there is a higher rate of aortic aneurysm or dissection among people who have relatives who have experienced this condition, which suggests a genetic link.

Treatment

- Acute
  - Time is essential
  - EKG
  - TEE
  - ST-segment elevation suggests of myocardial infarction should be treated as a primary cardiac event without delay.
  - CPT code 30130
  - STE
- Meds
  - Increasing pressure- control heart rate and blood pressure.
  - In patients with severe contraindications to beta-blockers, nondihydropyridine calcium channel blocking agents should be utilized as an alternative for heart control.
- Definitive Management
  - Urgent surgical consultation should be considered if there is a high degree of suspicion of thoracic aortic dissection regardless of the anatomic location (ascending versus descending), as soon as diagnosis is made or highly suspected.
  - Acute thoracic aortic dissection involving the ascending aorta should be urgently evaluated for emergent surgical repair because of the high risk of complications such as rupture.
  - Acute thoracic aortic dissection involving the descending aorta should be managed medically unless life-threatening complications develop (eg, malperfusion syndrome, progression of dissection, enlarging aneurysm, instability).

Treatment and Management

- If the aortic diameter increases rapidly or reaches a critical size (between 4.5-5.0 cm), elective surgery is recommended to prevent rupture. This involves surgically replacing the damaged portion of the aorta and sometimes repairing or replacing the aortic valve. Electrolyte surgery, prior to a dissection, is usually very successful in centers where there is experience with the procedure.
- In addition to absolute size, the rate of growth of the aorta, function of aortic valve and family history of dissection are used to determine timing of elective repair.
- Aortic dissections require immediate medical attention. In some instances, management with medication is possible. Surgery may be required depending on the location and the medical circumstances.
- Emergency surgery is required for ascending aortic dissections (Type A or proximal dissections). Descending aortic dissections may require emergency surgery. Patients must be transferred and evaluated at a tertiary hospital capable of performing the surgery. If the aorta ruptures as a result of an aneurysm or dissection, it is a life-threatening problem that requires immediate surgery.
Management

- Families with a genetic history of aortic aneurysms or dissections, it is recommended that family members at risk have their aortic root (first part of the ascending aorta) monitored by echocardiography (picture using sound waves) to evaluate the size of the root and how the aortic valves are working and the parts of the aorta further from the heart, specifically the aortic arch and descending aorta monitored by CT scan or MRI at least once a year.
- Once an aortic aneurysm or dissection is detected, imaging may be performed more often. The frequency will be based on several parameters and will be different for each patient. For example, more frequent monitoring is required if the aneurysm continues to increase in size, if rapid growth is observed, if aneurysm approaches critical sizes, if aortic valve function is compromised or if the individual’s aorta is approaching a size at which there is a family history of aortic dissection or rupture.
- Because aortic dissection occurs in a subset of individuals with bicuspid aortic valves, yearly monitoring by echocardiography to assess valve function and aortic root diameter is recommended.
- Medication, such as beta-blockers, to lower blood pressure and decrease the forcefulness of the heartbeat are often recommended to slow down enlargement or to hopefully prevent dissection of the aorta. Treatment with calcium channel antagonists should be considered in individuals who cannot tolerate beta-blockers.

Other Faces of Fatalities

Tyler Kahle

John Ritter

Whats Being Done

- TAD Coalition: www.tadcoalition.org
- www Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease
- A pocketbook of Guidelines for the Diagnosis and Management of Patients with Thoracic Aortic Disease is available free from the TAD Coalition
- Ritter Rules- combine knowledge with action
  - Life-saving guidelines to recognize, treat and prevent thoracic aortic dissections, a deadly tear in the large artery that carries blood away from the heart. Named for actor John Ritter, who died of a thoracic aortic dissection. Know the urgency, symptoms, who is at risk.
  - John Ritter Foundation is dedicated to promoting knowledge of aortic aneurysm and dissection through research and education. Its goal is to enlighten the general public while creating an open exchange of information at the highest levels of cardiovascular expertise. Its mission is to raise awareness of Aortic Disease, its detection, treatment and genetic predisposition.
- Pulitzer Prize-winning series on Aortic Aneurysm and Dissection: Kevin Helliker and Thomas M. Bynum, reporters at The Wall Street Journal, were awarded the 2004 Pulitzer Prize for Explanatory Reporting for uncovering and explaining the surprising prevalence of aneurysms.
Continued

- "Aortic Dissection at Any Age: The Tyler Kahle Story"
  Nebraska Methodist Health System is working to raise awareness and help healthcare providers understand and act on the knowledge that aortic dissection can occur at any age. The video: [Click here], produced with the help of Tyler’s family, offers educational information for healthcare providers and the general public.

- "Emergency Diagnosis and Treatment of Aortic Dissection": The NMF, in partnership with the New York State Department of Health, has created a comprehensive set of resources for emergency healthcare providers, including physicians, nurses, EMTs, and hospital risk managers. Available free to qualified healthcare professionals on the NMF website.

- Iowa Healthcare Collaborative Aortic Dissection Toolkit
  The NMF, in partnership with the New York State Department of Health, has created a comprehensive set of resources for emergency healthcare providers, including physicians, nurses, EMTs, and hospital risk managers. Available free to qualified healthcare professionals on the NMF website.

- Ehlers-Danlos Syndrome Network C.A.R.E.S. Inc.
  - Poster created to encourage awareness of vEDS, features photos of affected young people and list of common physical features of vEDS.
  - Genetic Testing at the Department of Medical Genetics at the University of Texas Medical School at Houston

October 2012

Conclusion

- Learning Experience
- Speak Up
- Use your resources
- 100%
- Make the step!!!!!!!

Resources

- Thoracic Aortic Disease Coalition
  (http://www.tadcoalition.org/tad/)
- Circulation: Guidelines for the diagnosis and Management of Patients with Thoracic Aortic Disease (AHA)
Thank you for Listening

IN LOVING MEMORY OF...

ADAM CARABAJAL
October 4, 1979 - July 19, 2009