

Page 1: Joint Announcement by AHA and ACC

Page 3: TAD Coalition Announced

Page 6: Ritter Rules

New Guidelines Aim to Prevent Unnecessary Death from Thoracic Aortic Disease

Multidisciplinary team of experts weighs in on diagnosis and management

When actor John Ritter died suddenly in 2003 from a tear in his thoracic aorta—the large artery that carries blood from the heart to the rest of the body—that tragedy brought attention to a rare but deadly condition that takes the lives of an estimated 10,000 Americans each year. Now, new clinical guidelines spearheaded by the American College of Cardiology (ACC) and the American Heart Association (AHA) not only offer new recommendations for the diagnosis and management of thoracic aortic disease (TAD), they deliver a powerful message to physicians and patients: Early diagnosis and treatment can save lives.

“If thoracic aortic disease can be detected early and managed, it gives us the opportunity to select patients for surgical or endovascular repair when the patient is stable,” said Loren F. Hiratzka, M.D., who chaired the guidelines writing committee and is the medical director of cardiac surgery for TriHealth, Inc. (Bethesda North and Good Samaritan Hospitals) in Cincinnati, OH. “The results of treatment for stable disease are far better than for acute—and often catastrophic—aortic rupture or dissection.”

The new guidelines appear in the April 6, 2010, issues of the *Journal of American College of Cardiology (JACC)* and *Circulation: Journal of the American Heart Association*, as well as on web sites of the ACC (www.acc.org) and the AHA (www.americanheart.org). They were developed in collaboration with the American Association for Thoracic Surgery (AATS), American College of Radiology (ACR), American Stroke Association (ASA), Society of Cardiovascular Anesthesiologists (SCA), Society for Cardiovascular Angiography and Interventions (SCAI), Society of Interventional Radiology (SIR), Society of Thoracic Surgeons (STS), and Society for Vascular Medicine (SVM). The American College of Emergency Physicians (ACEP) and the American College of Physicians (ACP) were also represented on the writing committee.

Recent scientific and clinical advances drove the development of guidelines to aid physicians in the diagnosis and management of aortic dissection, aortic aneurysm and other forms of TAD, said Kim A. Eagle, M.D., director of the University of Michigan Cardiovascular Center in Ann Arbor and co-author of the guidelines.

“We now have a deeper understanding of the genetic underpinnings of TAD, and we continue to expand our knowledge in this area,” he said. “There have been rapid advances in noninvasive imaging. Medical therapy is much better. Open surgical techniques with anesthesia have improved dramatically. We can even use endovascular (minimally invasive, catheter-based) approaches in some patients.”

An aortic aneurysm occurs when a portion of the aorta balloons out, increasing the diameter of the blood vessel by at least 50 percent at that spot. Although the wall of the aorta can become

dangerously thin, patients with an aortic aneurysm often have no symptoms unless the aneurysm ruptures.

In the case of aortic dissection, a tear in the inner lining of the aorta (the intima) allows blood to invade the middle layer (the media), creating a false passageway through which blood can flow. This false passageway steals a portion of the blood supply from the rest of the body. Classical symptoms include the sudden onset of intense pain in the chest, back, shoulder or abdomen. However, patients often experience less definite symptoms, which makes diagnosis difficult.

In aortic rupture, all three layers of the aortic wall burst, resulting in massive bleeding inside the body.

Risk factors for TAD include poorly controlled high blood pressure, advancing age, male gender, atherosclerosis, inflammatory diseases that damage the blood vessels, and certain genetic conditions that weaken connective tissue, such as Marfan syndrome. In addition, people whose aortic valve has only two leaflets (bicuspid valve) instead of the normal three leaflets may be at increased risk for an aortic aneurysm. Pregnancy, intense weight lifting and cocaine use increase the risk of aortic dissection.

One of the most important messages in the guidelines is that TAD often runs in families. As a result, family history is a critical tool for uncovering undiagnosed cases of TAD. Patients should tell their physicians not only about close relatives with aortic aneurysm, dissection, or rupture, but also about any family history of unexplained sudden death. “Family history is very important,” Dr. Eagle said. “Sudden cardiovascular collapse could have been a heart attack, but it could also have been sudden catastrophic aortic dissection.”

Additional highlights from the TAD guidelines include:

- Imaging of the thoracic aorta by computed tomography (CT), magnetic resonance imaging (MRI) or, in some cases, echocardiography is the best way to detect TAD and determine future risk. A chest x-ray alone is not sufficient.
- Patients with genetic conditions that increase the risk of TAD should have aortic imaging at the time of diagnosis to establish the size of the aorta, with periodic follow-up imaging thereafter.
- All patients with a bicuspid aortic valve should be evaluated to determine whether the aorta is dilating, or widening.
- The symptoms of acute aortic dissection, which can mimic those of a heart attack or another cause of chest pain, often make it difficult to arrive at a prompt diagnosis and may delay life-saving treatment. Physicians should keep aortic dissection in mind when asking questions about medical history, family history, and the type and pattern of pain, and when examining the patient.
- Aortic dissection involving the ascending aorta (the portion nearest the heart) is a life-threatening emergency that should be treated surgically.
- Aortic dissection involving the descending thoracic aorta may often be managed with medications that control the blood pressure and heart rate, unless life-threatening

complications develop. Additional medical therapy may include statins to lower elevated blood cholesterol levels.

- Minimally invasive endovascular techniques are an option in some patients with aneurysm or dissection of the descending thoracic aorta.
- All immediate relatives of a patient with thoracic aortic aneurysm or dissection, or a bicuspid aortic valve, should be evaluated by a cardiovascular physician and undergo aortic imaging to measure the size of the aorta and identify asymptomatic disease.

Not all health insurers pay for aortic imaging in high-risk asymptomatic patients, particularly based on family history, Dr. Hiratzka said.

“I hope the new guidelines will change that,” he said. “It could be lifesaving.”

“People with aortic disease do not have to die prematurely; they can live a long lifespan if they are diagnosed and receive treatment,” said Carolyn Levering, president and chief executive officer of the National Marfan Foundation, which convened the TAD (Thoracic Aortic Disease) Coalition of nonprofit, patient and professional groups. “That’s why the TAD Coalition has come together to launch a comprehensive public and medical awareness campaign to help maximize the impact of the new guidelines. Our first initiative is the dissemination of Ritter Rules, named to honor John Ritter. The purpose of Ritter Rules is to help people remember the important facts about aortic dissection so they can avoid the same kind of tragedy that took the life of the beloved actor.”

New Coalition Launches Multi-Faceted Campaign to Maximize Impact of New Aortic Disease Diagnosis and Treatment Guidelines from American Heart Association, American College of Cardiology and Nine Other Professional Organizations

TAD Coalition Announces “Ritter Rules” to Heighten Awareness of Risk Factors, Symptoms, Urgency and Diagnosis of Aortic Dissection

PORT WASHINGTON, NY – March 16, 2010 – Death from aortic dissection. It happens to the famous – actor John Ritter; RENT composer and lyricist Jonathan Larson; U.S. Olympic volleyball star Flo Hyman – and to the not famous – Tyler Kahle, 19, a loving son and brother; Paul Marks, 31, a loving husband and father; Allison Dunvegan Reed, 23, a loving daughter.

Now, as the country’s leading medical associations unveil diagnosis and treatment guidelines for thoracic aortic disease¹, the recently established Thoracic Aortic Disease (TAD) Coalition is launching a multi-faceted public and medical awareness campaign to maximize the impact of the guidelines and reduce the number of deaths from aortic dissection and rupture.

¹ Hiratzka LF, Bakris GL, Beckman JA, Bersin RM, Carr VF, Casey DE Jr, Eagle KA, Hermann LK, Isselbacher EM, Kazerooni EA, Kouchoukos NT, Lytle BW, Milewicz DM, Reich DL, Sen S, Shinn JA, Svensson LG, Williams DM. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. *Circulation* 2010; published online before print March 16, 2010, 10.1161/CIR.0b013e3181d4739e.

Today, the Coalition announced the creation of “Ritter Rules,” named to honor John Ritter. The purpose of Ritter Rules is to help raise awareness among the public about aortic dissection so they can reduce their risk of the same kind of tragedy that took the life of the beloved actor. Ritter Rules are not a part of the American Heart Association/American College of Cardiology guidelines, but rather an information tool the Coalition developed for the public.

“No one has to die or lose a loved one to aortic disease. Ritter Rules have been created as a tribute to John. His death brought renewed focus and a heightened awareness to the tragedy of aortic disease. With Ritter Rules, people at risk will be informed about their health and be able to advocate for their families and themselves,” said Amy Yasbeck, Ritter’s widow and founder of The John Ritter Foundation for Aortic Health.

Ritter Rules can be found on the TAD Coalition’s new website, www.TADCoalition.org. In addition, people can read basic information about aortic disease, learn about risk factors, read profiles of people who have suffered from aortic dissection, and find links to a wealth of resources to find out more. A link to the AHA/ACC guidelines is also available on the website.

TAD Coalition

The TAD Coalition, which was convened specifically to promote the new guidelines and help assure that people with aortic disease can get the highest quality of patient care, has three primary goals:

- Increase public awareness of both genetic and environmental factors that put people at risk of thoracic aortic aneurysms and acute aortic dissections
- Provide educational materials concerning the symptoms, clinical evaluation and medical management of thoracic aortic disease
- Improve the diagnosis and treatment of acute aortic dissections in the hospital emergency department

“The TAD Coalition is playing a critical role in helping to maximize the impact of the new thoracic aortic disease diagnosis and management guidelines,” said Loren Hiratzka, MD, who chaired the writing committee for the American College of Cardiology and the American Heart Association. “While these new guidelines will inform and update the medical community, the TAD Coalition will inform the public of risk factors and symptoms for these aortic diseases. Together, we hope there will be better outcomes and, certainly, a reduction in the number of deaths from aortic dissection and rupture due to earlier diagnosis and improved medical management.”

Members of the TAD Coalition include the Ehlers-Danlos National Foundation, GenTAC (Registry for Genetically Triggered Aortic Aneurysm and Dissection), International Registry of Acute Aortic Dissection (IRAD), John Ritter Foundation for Aortic Health, Loeys-Dietz Syndrome Foundation, National Marfan Foundation, Nebraska Methodist Health System and the Specialized Center of Clinically Oriented Research (SCCOR) on Thoracic Aortic Aneurysms and Dissections. The American Heart Association and the American College of Cardiology have partnered with the TAD Coalition to promote awareness and adoption of the new guidelines.

Critical Issues in Thoracic Aortic Disease

“We have been concerned for a long time about the frequency of deaths due to undiagnosed aortic disease leading to aortic dissections,” said Carolyn Levering, President and CEO of the National Marfan Foundation, which convened the TAD Coalition. “Now we are able to combine our efforts with those of other like-minded organizations, and with the backing of the most well-respected medical groups, bring extensive awareness of this condition to the general public and reinforce among the medical community the best diagnosis and treatment practices for thoracic aortic aneurysms and dissections. Together, we hope to prevent tragedy and loss.”

The critical issues in diagnosis and treating aortic disease, as outlined in the guidelines, include:

- Thoracic aortic diseases often have no symptoms and are not easily detectable until an acute and often catastrophic complication occurs.
- Only with specific imaging techniques can aortic disease be identified before a tear or rupture, yet some of the imaging techniques pose their own challenges, ranging from potential health risks to costs, which are not always covered for asymptomatic patients, even if they are deemed high risk.
- The urgency of treating stable, high risk patients, despite being asymptomatic, because the surgery has better results before an acute or catastrophic dissection or rupture occurs.
- Patients who are experiencing an aortic tear or rupture may have atypical symptoms, thus delaying the immediate care they need.
- There is a growing body of evidence that genetic changes or mutations predispose some people to aortic diseases. Therefore, identification of the genetic alterations leading to these aortic diseases has the potential for early identification of individuals at risk. Understanding the molecular basis may lead to targeted medical therapy to then treat the disease.
- More research is needed on aortic disease, its various causes and potential treatments.

Risk Factors

The primary risk factors for thoracic aortic disease are:

- Genetic syndromes, such as Marfan syndrome, Loeys-Dietz syndrome, Turner syndrome and vascular Ehlers-Danlos syndrome.
- Family history of thoracic aortic aneurysm and dissections, in which there is an inherited predisposition for aortic disease but the gene defect does not cause a specific syndrome. The genes that lead to non-syndromic forms of aortic aneurysm and dissections are in the early stages of identification.

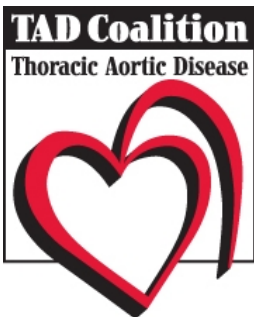
Other cardiovascular conditions associated with thoracic aortic aneurysm and dissection include bicuspid aortic valve and associated congenital variants in adults, and inflammatory diseases, such as Takayasu's arteritis and Behçet's disease.

Many people do not know that they have a genetic predisposition or a cardiac abnormality that would put them at an increased risk for thoracic aortic disease.

In addition, there are a number of conditions associated with increased aortic wall stress than can lead to an aortic dissection. These are:

- Hypertension, especially if it is uncontrolled
- Pheochromocytoma (a rare tumor of the adrenal gland)
- Cocaine or other stimulant use
- Weight lifting
- Trauma
- Deceleration or torsional (twisting) injury (motor vehicle accident, fall)
- Coarctation of the aorta (narrowing of the aorta)
- Pregnancy

For more information about thoracic aortic disease, please visit the TAD Coalition at www.TADCoalition.org.



Ritter Rules

Ritter Rules are life-saving reminders to recognize, treat and prevent thoracic aortic dissection, 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, Ritter Rules combine knowledge with action. Know the urgency, symptoms, who is most at risk and which imaging tests are required to diagnose this medical emergency.



URGENCY: Thoracic aortic dissection is a medical emergency. The death rate increases 1% every hour the diagnosis and surgical repair are delayed.

PAIN: Severe pain is the #1 symptom. Seek immediate emergency medical care for a sudden onset of severe pain in the chest, stomach, back or neck. The pain is likely to be sharp, tearing, ripping, moving or so unlike any pain you have ever had that you feel something is very wrong.

MISDIAGNOSIS: Aortic dissection can mimic heart attack. Heart attacks are far more common than aortic dissection. But if a heart attack or other important diagnosis is not clearly and quickly established, then aortic dissection should be quickly considered and ruled out, particularly if a patient has a family history or features of a genetic syndrome that predisposes the patient to an aortic aneurysm or dissection.

IMAGING: Get the right scan to rule out aortic dissection. Only three types of imaging studies can identify aortic aneurysms and dissections: CT, MRI and transesophageal echocardiogram. A chest X-ray or EKG cannot rule out aortic dissection.

RISK FACTOR: Aortic dissections are often preceded by an enlargement of the first part of the aorta where it comes out of the heart, called an aortic aneurysm. If you have an aneurysm, you are at increased risk for an aortic dissection.

RISK FACTOR: A personal or family history of thoracic disease puts you at risk. If you or a family member is living with an aneurysm or if you have a family member who has had an aortic dissection, you are at an increased risk for thoracic aortic dissection. You and your other family members should be evaluated to determine if a predisposition for aortic aneurysm and dissection is running in the family.

RISK FACTOR: Certain genetic syndromes put you at risk. These genetic syndromes greatly increase your risk for thoracic aortic disease and a potentially fatal aortic dissection: Marfan syndrome, Loeys-Dietz syndrome, Turner syndrome and vascular Ehlers-Danlos syndrome.

RISK FACTOR: Bicuspid aortic valve disease puts you at risk. If you have a bicuspid aortic valve (two leaflets instead of the typical three), or have had a bicuspid aortic valve replaced, you need to be monitored for thoracic aortic disease.

TRIGGERS: Lifestyle and trauma can trigger aortic dissection. It is possible to trigger an aortic dissection through injury to the chest, extreme straining associated with body building, illicit drug abuse, poorly controlled high blood pressure or by discontinuing necessary blood pressure medications. Rarely, pregnancy can trigger an aortic dissection. However, women with aortic aneurysms and connective tissue disorders who are pregnant are at higher risk of aortic dissection during late pregnancy and delivery and should be carefully monitored by a cardiovascular specialist.

PREVENTION: Medical management is essential to preventing aortic dissection. If you have thoracic aortic disease, medical management that includes optimal blood pressure control, aortic imaging and genetic counseling is strongly recommended. Talk with your physician.