Rev Bras Cardiol Invasiva.
2014;22(3):303-7

Case Report

Atypical Presentation of Intramural Hematoma of the Ascending Aorta Using a Conservative Approach

Claudia Maria Rodrigues Alves[1,2], Manuel Pereira Marques Gomes Jr.[1,2], Ricardo Peressoni Faraco[1,2], Tatiana Sawabini[2], Paulo Cézar Ferraz Dias Filho[2], Hilton Muniz Leão Filho[2]

ABSTRACT

Aortic intramural hematoma is a severe disease, secondary to *vasa vasorum* bleeding and/or aortic micro-ulcers without connection with the true lumen of the vessel. Its occasional finding in the absence of symptoms or aortic trauma is quite rare. In this report, an asymptomatic female patient with involvement of the ascending aorta and transverse arch was maintained on clinical treatment, despite the prevailing recommendations for surgical repair in Stanford type A lesions. Relevant aspects of lesion characterization in different imaging tests, identification of high risk anatomical criteria and conduct are discussed. A satisfactory 7-month follow-up was observed.

Descriptors: Aorta, thoracic. Aortic diseases. Hematoma.

RESUMO

Apresentação Atípica de Hematoma Intramural da Aorta Ascendente com Tratamento Conservador

O hematoma intramural aórtico é uma doença grave, secundária a sangramento da *vasa vasorum* e/ou microúlceras aórticas, sem comunicação com a luz verdadeira do vaso. Seu achado ocasional, na ausência de sintomas ou trauma aórtico, é bastante raro. Neste relato, paciente assintomática, com comprometimento da aorta ascendente e arco, foi mantida em tratamento clínico, a despeito das recomendações predominantes para correção cirúrgica em lesões classificadas como tipo A de Stanford. Discutimos aspectos pertinentes à caracterização da lesão nos diferentes exames de imagem, identificação de critérios de alto risco anatômico e condução do caso. A evolução em 7 meses foi satisfatória.

Descritores: Aorta torácica. Doenças da aorta. Hematoma.

Since the original description of acute aortic syndromes,[1] the growing diagnostic readiness and the availability of sophisticated imaging studies have allowed an early identification of patients with acute and typical aortic scenarios. In the case of intramural hematomas (IMH), as occurs in classic cases of dissection, the agility in diagnosis and the immediate treatment, especially for lesions classified as type A, are also crucial to success. A more widespread recognition of these diseases and the frequent use of advanced diagnostic methods can promote a clinical situation in which the image outweighs the importance or clinical significance of the disease itself. In this study, the case of an asymptomatic female patient presenting IMH of ascending aorta is described, with emphasis on diagnostic imaging and on medical management.

CASE REPORT

A female patient, aged 49, white, with a history of controlled hypertension and glucose intolerance, underwent preoperative evaluation for a hysterectomy for uterine myomatosis, during which an angiotomography of coronary arteries was requested, due to a family history of early coronary disease. In this procedure, performed on an outpatient basis, the presence of IMH of ascending aorta and transverse arch was observed as an incidental finding, occupying the entire circumference of the aorta, with a maximum thickness of 8 mm and discrete proximal progression to cervical vessels. Furthermore, a slight compression of the right coronary artery ostium was noticed (Figure 1).

The patient was referred for hospital admission and medical management. In a previous clinical history, she related an episode of prolonged fever that had occurred 16 years ago, accompanied by pleural effusion, when an investigation for systemic lupus erythematosus and tuberculosis (sic) was performed and subsequently discarded.

Since then, the patient was asymptomatic and practicing regular exercise (cycling); the woman denied

[1] Escola Paulista de Medicina, Universidade Federal de São Paulo, São Paulo, SP, Brazil.

[2] Hospital do Coração Associação do Sanatório Sírio, São Paulo, SP, Brazil.

Correspondence to: Claudia Maria Rodrigues Alves. Rua Simão Álvares, 527/63 – Pinheiros – CEP: 05417030 – São Paulo, SP, Brazil
E-mail: cmralves@uol.com.br

Received on: 06/12/2014 • Accepted on: 08/28/2014

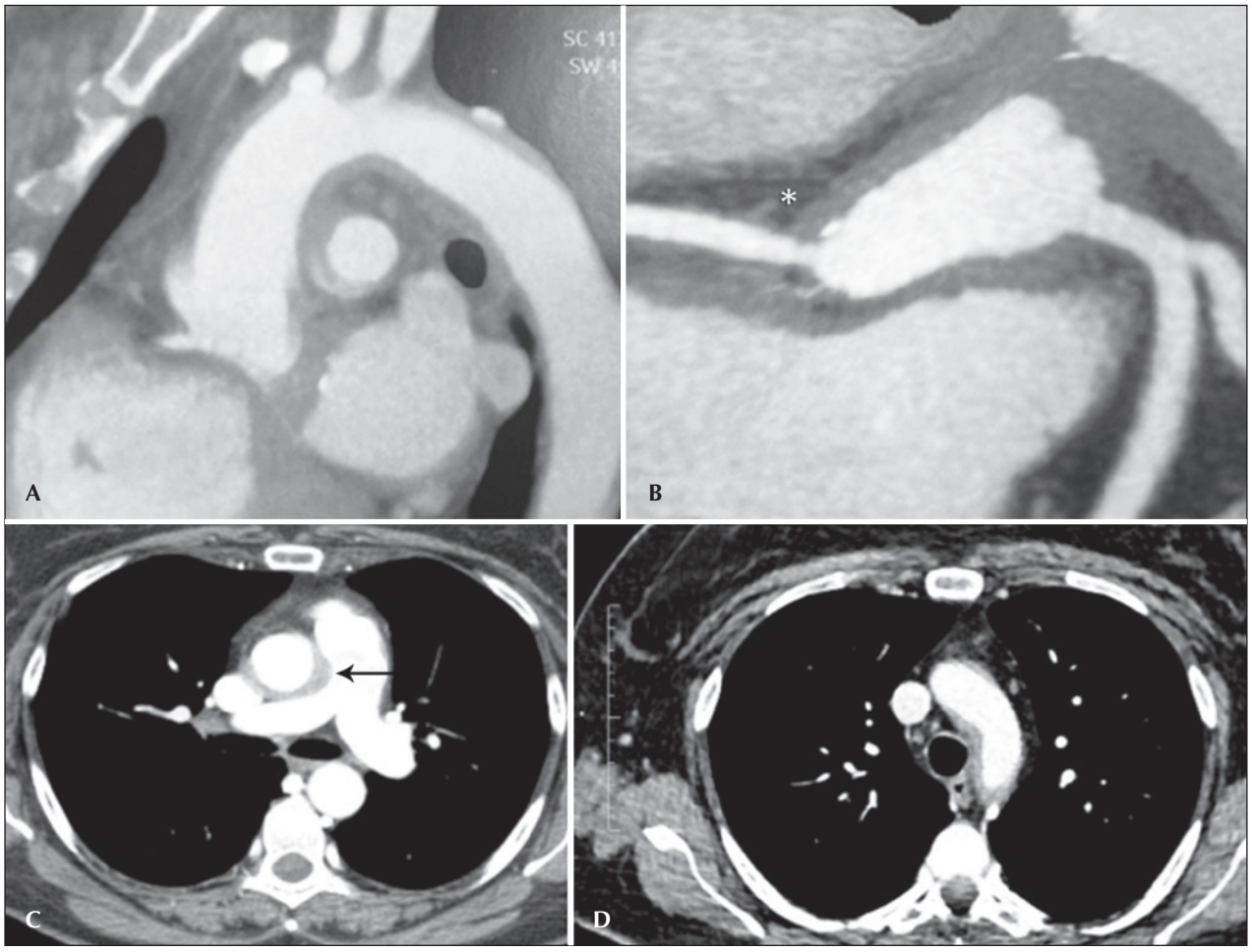

Figure 1 – Multi-row detector computerized tomography angiography. There is a clear circumferential aortic involvement extending into the transverse arch, with involvement of the initial portion of cervical vessels (A) and right coronary artery ostium (B*). In cross-section, a slight pulmonary artery compression can be observed in the thickest point of the intramural hematoma, extending intothe aortic arc (D).

previous episodes of chest pain or chest trauma at any time. Furthermore, no manifestation compatible with systemic inflammatory disease was perceived. The patient denied any family history of aortic disease or sudden death.

At the time of hospital admission, laboratory exams revealed metabolic profile, serum protein electrophoresis, and inflammatory activity profile (C-reactive protein, erythrocyte sedimentation rate, serum complement, rheumatoid factor, and antinuclear antibodies) with in normality; human immunodeficiency virus and syphilis serology were also negative. The hemogram revealed anemia (hemoglobin = 10.3 g/dL), which was considered compatible with her long-term hypermenorrhea. The electrocardiogram showed first-degree atrioventricular block, and an echocardiogram revealed a mild thickening of aortic valve, with a mean gradient of 8 mmHg.

Considering that the patient was clinically stable, she was maintained under close observation in the hospital, and a first control with computerized tomography angiography (CT-A) of thoracic aorta was scheduled for the fifth day of hospitalization, in association with positron emission tomography (PET-CT), which demonstrated stability in aortic diameter (maximum diameter = 34 mm), predominance of the hematoma in posterolateral portions to the left of the ascending aorta, and with an apparently slight reduction of the hematoma, but with maintenance of a thickness of 8 mm. The pulmonary artery diameter was normal. In PET-CT, fluoro-deoxyglucose (FDG) uptake was observed in the portion corresponding to the highest volume of the hematoma (maximum standard uptake value – SUV max = 4.4), without abnormal uptake in other parts of the aorta or organs (Figure 2).

The patient was discharged, having been told to avoid contact sports or resistance exercises, and was prescribed beta blocker, angiotensin receptor blocker, and statin. At that time, a new control was scheduled to be held in 15 to 20 days, if the patient remained

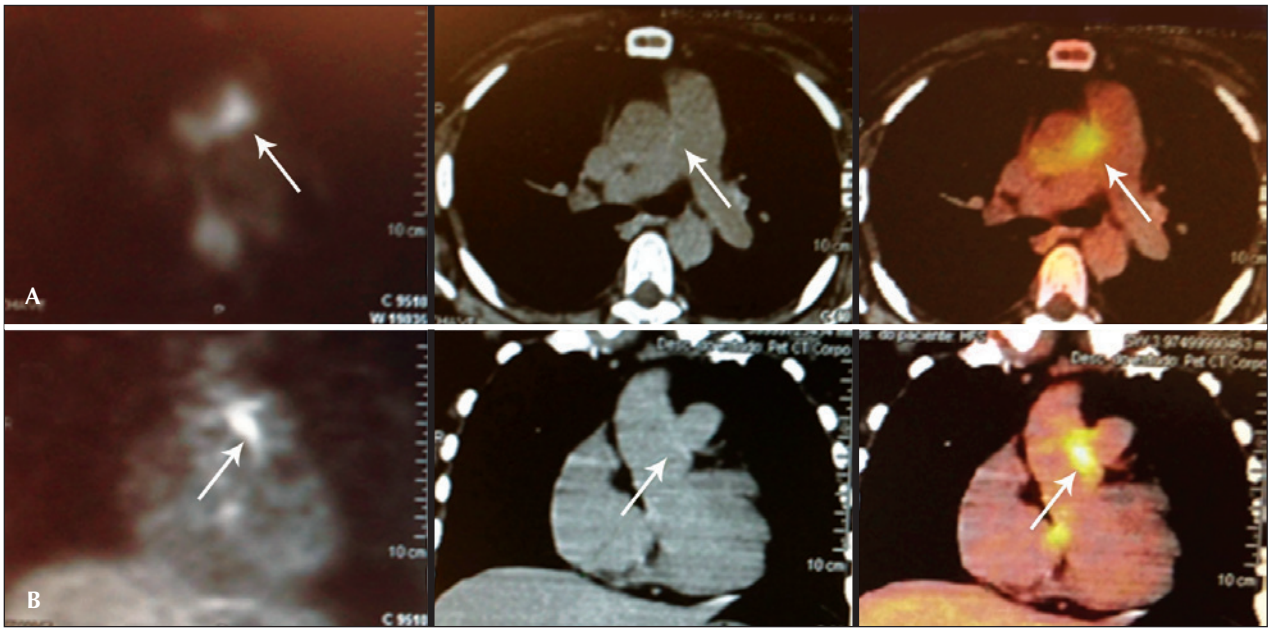

Figure 2 – Positron emission tomography associated with computed tomography (PET-CT). Cross-sections at the level of pulmonary artery bifurcation (A) and sagittal sequences (B) show spontaneously hyperdense, circumferential thickening of ascending aorta, with signs of increased glucose metabolism (SUV max = 4.4).

asymptomatic. This third CT-Acontrol revealed hematoma stability.

After seven months, the patient remained asymptomatic and in regular use of the above-mentioned drugs and a new CT scan of thoracic aorta revealed minimal reduction in IMH thickness (7 mm) and maintenance of aortic diameter (Figure 3).

DISCUSSION

Corresponding to about 6 to 10% of cases of acute aortic syndrome, IMH of ascending aorta diagnosed during the investigation of an episode of acute chest pain is a severe disease with high mortality.[2] The possibility of progression to dissection or to frank rupture (20 to 50%) underlies the indication of immediate surgical treatment for a great number of patients, particularly those whose injuries are situated in the ascending aorta. However, in recent years, several studies – especially in the Asian population – have favored a strategy of close observation for stable patients without signs of poor prognosis, e.g., persistent pain, aortic diameter increased on admission (> 48-55 mm), hematoma thickness > 11 mm, a concomitant presence of ulcer-like images (not necessarily characteristic of a penetrating ulcer), or hematoma progression.[3] Thus, in stable patients, the follow-up with imaging studies is a mandatory step in the monitoring and in treatment indication.

Asymptomatic IMH is rarely described in the literature (two cases). One patient exhibited progression to a penetrating ulcer and underwent surgical correction,[4] and another patient with vague symptoms underwent an emergency thoracotomy, during which an aortitis accompanied by wall thickening was diagnosed,[5] without replacement of the aortic segment affected.

Despite the extent of IMH found in this case and not withstanding its classification (Stanford type A), the option in favor of a conservative management was based on an absence of clinical manifestations of the disease and on CT criteria for high risk, weighed against the morbidity and mortality inherent to an extensive aortic surgery.

In the case of an asymptomatic patient undergoing CT or PET-CT for any indication (coronary heart disease investigation, cancer monitoring, etc.) and in whom IMH was detected, it is critical to be alert to the possibility of differential diagnoses. In the present case, images synchronized with the electrocardiogram were requested in order to exclude artifacts, and the possibility of aortitis was investigated, although, at first, the patient's clinical evaluation and laboratory tests were not suggestive of this hypothesis.

The characteristic finding of IMH is the presence of a thrombus (crescent-shaped, or occupying the entire aortic circumference) in the vessel wall, without communication with the vessel lumen, caused by *vasa vasorum* rupture or by the occurrence of undetected micro ruptures or micro ulcers. With multi-row detector CT-A, the wall thickening with high attenuation (60-70 Hounsfield Units – HU), observed in the unenhanced series, is clearly seen in the contrast-enhanced series

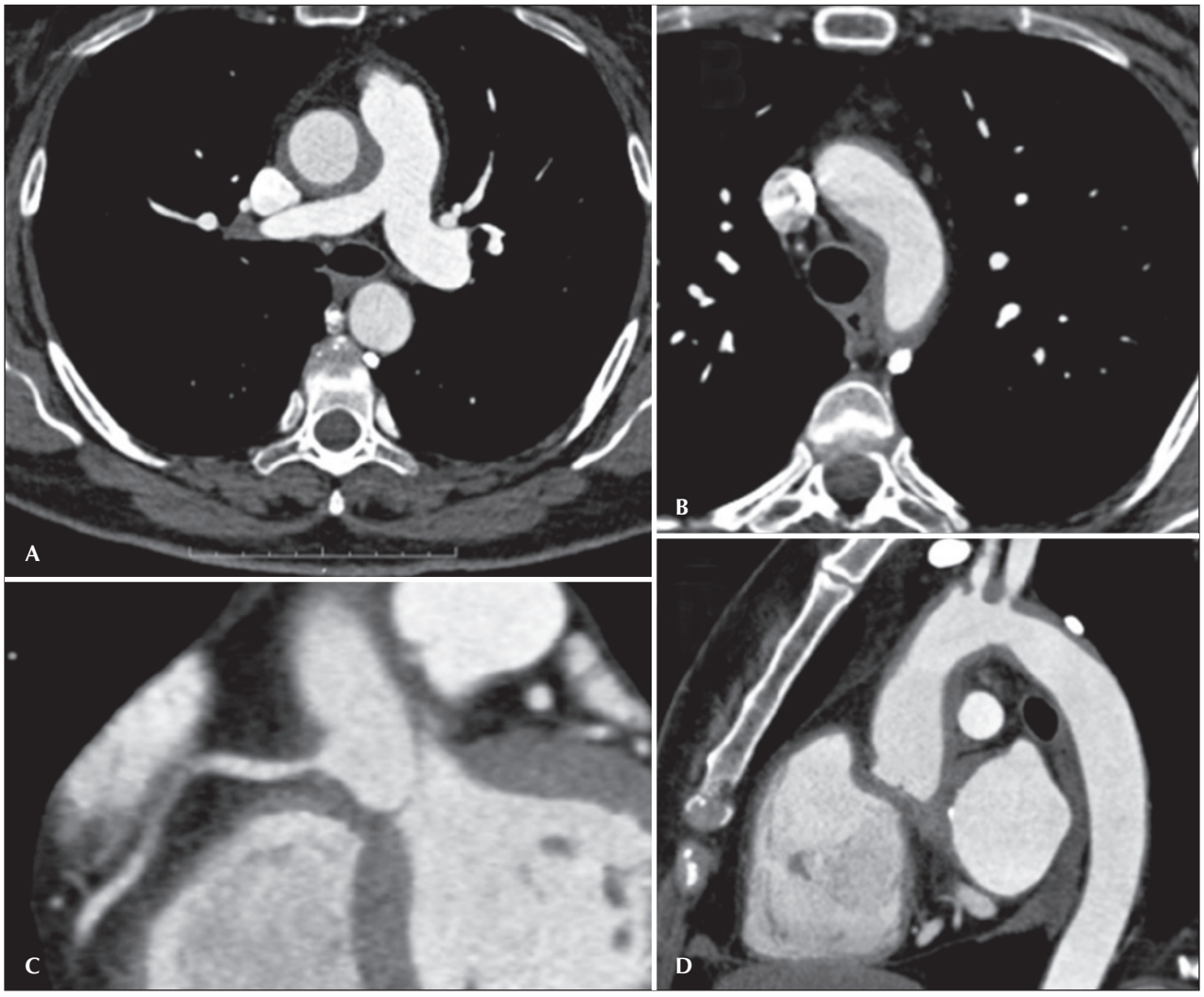

Figure 3 – Multi-row detector computerized tomography angiography performed 7 months after treatment. Although the measured thickness of the hematoma is slightly smaller, with an apparent reduction in hematoma volume and maintenance of aortic diameters, the values are still within the range of variation of the method (8 and 7 mm, initial and final). Transverse sequences at pulmonary artery bifurcation (A) and aortic arc (B) levels. Reconstruction at the level of right coronary artery outflow tract (C) and longitudinal section of aortic arch and great vessels (D).

as a separate image, with no communication with the lumen.[6] In MRI, additionally to the aforementioned anatomical aspect, a hyper-intense signal on T2-weighted sequences and an iso- or hyper-intense signal on T1-weighted sequences (black-blood), depending on the stage of evolution, are characteristic findings.[7]

In cases of aortitis, the characteristic finding of CT-A in the unenhanced phase, as opposed to hematomata, is the low wall attenuation (< 40 HU). Another picture that leads to confusion is the presence of an intraluminal mural thrombus, often not distinguishable by CT studies. In case of persistent doubt, the differentiation may be achieved with magnetic resonance imaging (MRI), due to the presence of a hypo-intense or iso-intense signal.[8]

While PET-CT is used predominantly in malignancy or inflammatory disease investigations, FDG uptake is also observed in cases of atherosclerosis and in the presence of a parietal thrombus.[4] In the present case, PET-CT revealed a concentrated hyper-uptake area at the point of greatest volume and excluded uptake occurrence in other aortic portions or organs. The use of PET-CT in the prognostic assessment of patients, correlating the marked uptake with an increased risk of expansion and rupture, is still contradictory, but this may be a useful marker.[9]

At this stage, no further investigation on myocardial ischemia was conducted, considering that the patient exhibited only mild compression in the right coronary

artery ostium, and also because the possibility of surgery was not excluded.

In the authors' opinion, the clinical and imaging follow-up was an appropriate conduct in this case, avoiding a large surgical procedure, and also considering that the control follow-up revealed IMH stability.

The main stays of conservative treatment are the knowledge of relevant data pertaining to the diagnostic investigation by imaging studies and the association of these findings with the clinical assessment. The rapid development of these imaging methods and the low frequency of aortic diseases in the cardiology practice require a multidisciplinary participation. It is required that radiology services (or, when necessary, the physician) are accustomed to deliver reports that include the largest amount of relevant information for an initial risk classification, even if imperfect, and that inform the attenuation values observed in unenhanced series. An appropriate and sufficient clinical care strategy includes the classes of drugs that probably play a role in the reduction of aortic events[10] (beta blockers, ACE inhibitors or angiotensin receptor blockers, and statins) and patient monitoring by imaging studies (preferably with nuclear magnetic resonance, to reduce exposure to X-ray). Considering that the pathophysiology of aortic diseases has only begun to be elucidated, it is possible to assume that silent episodes of intramural bleeding can occur as a mechanism of early dilation, or even of dissection. In this particular case, the possibility of progression to a late aortic dilation cannot be excluded.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

FUNDING SOURCES

None.

REFERENCES

1. Vilacosta I, Román JA. Acute aortic syndrome. Heart. 2001; 85(4):365-8.
2. Harris KM, Braverman AC, Eagle KA, Woznicki EM, Pyeritz RE, Myrmel T, et al. Acute aortic intramural hematoma: an analysis from the International Registry of Acute Aortic Dissection. Circulation. 2012;126(11 Suppl 1):S91-6.
3. Kruse MJ, Johnson PT, Fishman EK, Zimmerman SL. Aortic intramural hematoma: review of high-risk imaging features. J Cardiovasc Comput Tomogr. 2013;7(4):267-72.
4. Hayashida T, Sueyoshi E, Sakamoto I, Uetani M, Chiba K. PET features of aortic diseases. AJR Am J Roentgenol. 2010;195(1):229-33.
5. Yamane K, Hirose H, Youdelman BA, Diehl JT. Thickened ascending aortic wall mimicking intramural hematoma. Asian Cardiovasc Thorac Ann. 2012;20(2):208-9.
6. Hallinan JTPD, Anil G. Multi-detector computed tomography in the diagnosis and management of acute aortic syndromes. World J Radiol. 2014;6(6):355-65.
7. Hartlage GR, Palios J, Barron BJ, Stillman AE, Bossone E, Clements SD, et al. Multimodality imaging of aortitis. JACC Cardiovasc Imaging. 2014;7(6):605-19.
8. Baliga RR, Nienaber CA, Bossone E, Oh JK, Isselbacher EM, Sechtem U, et al. The role of imaging in aortic dissection and related syndromes. JACC Cardiovasc Imaging. 2014;7(4): 406-24.
9. Golestani R, Sadeghi MM. Emergence of molecular imaging of aortic aneurysm: implications for risk stratification and management. J Nucl Cardiol. 2014 ;21(2):251-67; quiz 268-70.
10. Hiratzka LF, Bakris GL, Beckman JA, Bersin RM, Carr VF, Casey DE Jr, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease: executive summary. 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. Catheter Cardiovasc Interv. 2010;76(2):E43-86.