Diagnosis and management issues in thoracic aortic aneurysm

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Thoracic aortic enlargement is an increasingly recognized condition that is often diagnosed on imaging studies performed for unrelated indications. The risk of unrecognized and untreated aortic enlargement and aneurysm includes aortic rupture and dissection which carry a high burden of morbidity and mortality. The etiologies underlying thoracic aortic enlargement are diverse and can range from degenerative or hypertension associated aortic enlargement to more rare genetic disorders. Therefore, the evaluation and management of these patients can be complex and requires knowledge of the pathophysiology associated with thoracic aortic dilation and aneurysm. Additionally, there have been important advances in the treatment available to patients with thoracic aortic disease, including an increased role of endovascular therapy. Given the risk of mortality, increased clinical recognition and advances in therapeutics, the American College of Cardiology, American Heart Association and related professional societies have recently published guidelines on the management of thoracic aortic disease. This review focuses on the pathophysiology and various etiologies that lead to thoracic aortic aneurysm along with the diagnostic modalities and management of asymptomatic patients with thoracic aortic disease. (Am Heart J 2011;162:38-46.e1.)

Thoracic aortic aneurysm

Thoracic aortic aneurysms (TAA) are diagnosed most commonly on an imaging study performed for an unrelated indication. Thoracic aortic dilation occurs with an estimated incidence of 5.6 to 10.4 cases per 100,000 patient-years and an estimated prevalence of up to 4.2% of the general population without hypertension.1,2 Dilation is defined as a measurement that exceeds the range of normal for a given age and body size. Aneurysm is defined as a 50% increase above this range. Clinically, TAA are typically silent until a complication presents. Rarely, depending on size and location, TAA can present with dysphagia, hoarseness, chest or back pain, signs, or symptoms of aortic insufficiency.3 Although a relatively rare process as compared to other cardiovascular diseases, the devastating consequences of untreated or undiagnosed TAA (ie, rupture, dissection) elevate its clinical importance.

Pathophysiology

The pathophysiologic basis for TAA is complex and related, in part, to underlying conditions that predispose patients to aortic dilation. Although the specific pathophysiologic pathways are still being assessed, a general scheme for explaining the clinical findings of thoracic aortic aneurysm include specific genetic mutations or predisposition leading to decreased contractile function of vascular smooth muscle cells. This process then leads to excess inflammatory mediators (transforming growth factor-B [TGF-B], penin-argrotensin-aldosterone system [RAS], and insulin-like growth factor-1 [IGF-1]) and increased activation of stretch pathways which, in turn, increase proteoglycan and matrix metalloproteinase production. This inflammatory milieu furthers tissue degradation and results in progressive aortic wall weakening and subsequent aortic dilation. Histologically, the pattern of cystic medial necrosis and loss of elastic fibers are commonly seen as the end result of this biochemical pathway.4

Natural history

In general, isolated idiopathic or degenerative ascending aortic aneurysms will have a rate of growth of ~0.1 cm/y. Natural history studies have shown that after an aneurysm reaches 6 cm in the ascending aorta and 7 cm in the descending aorta, the complication rates are greatly increased.5 In addition, if the aorta is >5 cm at the time of diagnosis, the growth rate may be accelerated, up to 0.1 to 0.15 cm/y. This more rapid increase in size increases the risk of aortic complications. Certain inherited conditions, discussed in detail below, that cause a predisposition to TAA may also contribute to accelerated growth.6

Etiology

Various etiologies of TAA are listed in Table 1. In older patients, the most common etiology of TAA is a degenerative process associated with increased age; however, hypertension, tobacco abuse, hyperlipidemia, and genetic factors can result in acceleration of growth.6
Genetically triggered thoracic aortic aneurysm syndromes

Marfan syndrome
- Most common inherited connective tissue disease
- Mutation in FBN-1 gene leads to decreased tensile strength of the aorta
- Estimated that 75% of patients will have a dilated aortic root

Loeys-Dietz Syndrome
- Aggressive vasculopathy linked to TGFBR1 or 2 mutation
- Early detection and intervention is important

Bicuspid aortic valve
- ≥50% have tubular/ascending aneurysm
- 20% sinus of Valsalva involvement
- Faster rate of growth than aneurysms associated with a three leaflet valve

Turners syndrome
- 1/3 with bicuspid valve and coarctation of the aorta
- Ascending aortic aneurysm

Familial non-syndromic thoracic aortic aneurysm syndrome
- Dilated aorta
- Absence of other connective tissue disease
- Family history dissection/aneurysm

Aortitis
- Infectious
  - Syphilis (historical)
  - Salmonella
  - Staphylococcal species
  - Mycobacterium
- Non infectious/inflammatory
  - More common:
    - Giant cell and Takayasu arteritis
  - Less common:
    - Behcets, Cogan’s syndrome, relapsing polychondritis
  - Rare:
    - Rheumatoid arthritis, spondyloarthropathies

Trauma
- Typical location is at the aortic isthmus
- Complications include rupture, pseudoaneurysm, chronic dissection with secondary aneurysm formation

Chronic aortic dissection
- Aneurysm due to growth and pressure differential of false lumen

The inheritance pattern in those with genetically triggered aneurysm syndromes is not fully elucidated but is thought to be autosomal dominant with reduced penetrance, which accounts for the phenotypic variability that can be seen in these cases. Nearly 20% of patients who develop a TAA will have a family history of aortic disease, and a familial pattern should be suspected in patients who present at a young age. These genetically triggered TAA tend have the most aggressive rate of growth and are more likely to require intervention.
root aneurysm, ectopic lentis, a pathogenic fibrillin-1 (FBN-1) mutation, or systemic features on physical exam. The absence of a family history requires the presence of an aortic aneurysm and one of the following: ectopic lentis, pathogenic FBN-1 mutation, systemic features, or aortic enlargement not meeting Z-score ≥2 with both ectopic lentis and FBN-1 mutation.

Typically, the aortic dilation seen in the Marfan syndrome is localized to the aortic root but may extend into the ascending aorta and is associated with an accelerated growth rate (up to 0.2-0.3 cm/y) as compared with degenerative aneurysms. Defects in the FBN-1 gene have been found to lead to excess TGF-β signaling and subsequent increased tissue degradation and weakening of the aortic wall.12,15 Histologically, the aortic tissue in patients with Marfan syndrome have a fraction of the elastin of normal aortas and increased elastin fragmentation.14 The end result of these processes predisposes patients with the Marfan syndrome to a high risk of aortic complications at a relatively young age. The criteria for operative repair in these cases is outlined below and is more aggressive than in patients with degenerative aneurysms. In many cases, the aortic valve is morphologically normal and may be able to be resuspended/reimplanted at the time of aortic repair.15,16

Vascular ED

Patients with vascular ED represent a rare but extremely high risk group. Vascular ED syndrome is an inherited connective tissue disease that results from a defect in type III procollagen due to mutations in the COL3A1 gene. Vascular ED syndrome affects 1 in 5 to 20,000 births. Vascular and connective tissue integrity is markedly impaired, and subsequently, these patients can have significant complications including gastrointestinal complications and uterine rupture in pregnancy. The vascular tissue in these patients is very weak and difficult to safely manipulate in the operating room, even in experienced hands. These patients require specialized care.17

<table>
<thead>
<tr>
<th>Table II. Imaging the thoracic aorta</th>
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<tbody>
<tr>
<td><strong>Modality</strong></td>
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<tr>
<td>MDCT angiography</td>
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<tr>
<td>MRI/MR angiography</td>
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<tr>
<td>Transesophageal echocardiography</td>
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Familial nonsyndromic thoracic aortic aneurysm syndromes

Familial nonsyndromic thoracic aortic aneurysm is a term applied to patients with a family and personal history of aortic aneurysm without meeting strict criteria of known syndromes. This syndrome can include patients with a dilated aorta and a family history of dissection or rupture or sudden unexplained death and are becoming increasingly recognized. Up to 20% of patients with an isolated thoracic aortic aneurysm will have a family member with a similar history.6 These patients tend to present at a younger age (mean 55 years) and have a faster rate of growth than degenerative/sporadic aneurysms (up to 0.2 cm/y). The ascending thoracic aorta is more commonly involved (~80%) than the descending aorta (~20%). The variable penetrance and expression of the identified mutations (ACTA 2, MYH11 [also associated with patent ductus arteriosis], and TGFBR2) make definitive diagnosis difficult.18,19 ACTA2 may account for up to 14% of genetic mutations associated with familial syndromes and testing for ACTA2 is a class IIa recommendation in the most recent guidelines for patients with a thoracic aortic aneurysm and a family history of thoracic aortic aneurysm and/or dissection.16 ACTA2 encodes smooth muscle α2 actin. The deficiency of smooth muscle α2 actin leads to decreased numbers and disorganization of
vascular smooth muscle cells with subsequent loss of elastic fibers leading to decreased adaptation to stress in aortic tissue and aneurysm formation. Bicuspid aortic valve

Bicuspid aortic valve (BAV) disease occurs in 1-2% of the general population and is the most commonly recognized congenital abnormality in adulthood. The association with BAV and aortopathy/aneurysm formation has been well established. Embryologically, the aortic valve and the ascending aorta share the same neural crest origin. Histologically, aortic tissue in BAV patients shows medial degeneration, increased matrix metalloproteinase activity and decreased FBN-1 in the aortic wall. Subsequent weakening of the aortic wall leads to accelerated growth rates for, BAV associated aneurysms, up to 0.2 cm/y. Aortic aneurysm associated with BAV can occur in up to 50% of cases and involve the root through the proximal arch. However, coarctation of the aorta is also highly associated with a BAV (up to 50% of coarctation patients will have a BAV), suggesting that the abnormal aortic tissue may extend into the distal arch or proximal descending aorta.

BAV is thought to be an inherited condition, and like the other inherited conditions discussed, is likely autosomal dominant inheritance with reduced penetrance. The heritable nature of BAV is supported by data that shows that even in first degree relatives of patients with BAV who have normally functioning three leaflet valve, there is evidence of aortopathy with increased size, less distensibility, and increased wall stiffness in the absence of a BAV. For this reason, screening of the aortic valve and proximal ascending aorta with echocardiography or another modality should be recommended for first-degree relatives of patients with BAV.

Overall, it is estimated that 25% of patients with a BAV will require an intervention to the valve, aorta, or both during their lifetime. BAV is also associated with a risk of endocarditis in up to 2% of cases. However, antibiotic prophylaxis before dental work or other procedures is not recommended in the most recent American College of Cardiology/American Heart Association guidelines on this topic.

Criteria for operative repair in cases of BAV associated aneurysm is outlined below and is dependent both on the size of the aneurysm and concomitant valvular dysfunction. However, given the elevated rate of aortic expansion in patients with BAV, early elective repair based on the size of the aneurysm alone may be indicated, irrespective of the valve disease severity. If aortic valve stenosis or insufficiency is the indication for intervention, the abnormal aortic tissue should also be addressed and potentially replaced at the time of valve surgery if the diameter exceeds >4 to 4.5 cm.

Loeys Dietz syndrome

Loeys Dietz is a recently described syndrome that involves defects in TGFBR1 or TGFBR2. Phenotypic characteristics include hypertelorism, cleft palate or split uvula along with some shared characteristics with the Marfan syndrome. Patients with this syndrome are at high risk at a young age for aortic dilation, rupture or dissection. For this reason, very early operative intervention is recommended at ascending aortic diameters of ≥4.2 cm by transesophageal echocardiography (TEE) or ≥4.4 to 4.6 cm by computed tomography (CT).

Initial diagnosis and subsequent workup

Once thoracic aortic dilation is suggested by any imaging modality, further workup and confirmatory imaging should be performed.

### Table III. Thoracic aortic aneurysm - chronic medical therapy

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dosage</th>
<th>Goal of therapy</th>
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</thead>
<tbody>
<tr>
<td><strong>First-line agent:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• β-Blockers</td>
<td>Titrate to effect</td>
<td>• Heart rate &lt;60 beat/min</td>
</tr>
<tr>
<td><strong>Second Line Agents:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Angiotensin Receptor Blockers</td>
<td>Start at a low dose and titrate to BP goals</td>
<td>• Blood pressure &lt;130/80 mm Hg</td>
</tr>
<tr>
<td>• ACE inhibitors</td>
<td></td>
<td>• Use in all pts with Marfan syndrome</td>
</tr>
<tr>
<td><strong>Third-line agent:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Calcium-channel blocker (dihydropyridine) Statins</td>
<td>Start at a low dose and titrate to above goals</td>
<td>• Goal LDL &lt;70 ng/dL if atherosclerosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• LDL &lt;100 ng/dL if no atherosclerosis</td>
</tr>
<tr>
<td><strong>Tobacco cessation aids</strong></td>
<td>Standard dosing</td>
<td>• Smoking cessation</td>
</tr>
<tr>
<td>• Varenicline</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Chantix</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Bupropion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Nicotine replacement</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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Patients undergoing aortic valve repair or replacement

Table IV. Size criteria for elective surgical intervention

<table>
<thead>
<tr>
<th>Condition</th>
<th>Location and size</th>
</tr>
</thead>
<tbody>
<tr>
<td>Degenerative thoracic aneurysm</td>
<td>Ascending aorta or arch ≥5.5 cm or Descending aorta ≥6 cm</td>
</tr>
<tr>
<td>Chronic aortic dissection</td>
<td>ASCENDING AORTA OR ARCH ≥5.5 CM OR DESCENDING AORTA ≥6 CM</td>
</tr>
<tr>
<td>Chronic traumatic dissection/ \n  Pseudoaneurysm</td>
<td>ASCENDING 4.0-5.0 CM OR DESCENDING ≥5.5 CM</td>
</tr>
<tr>
<td>Intramural hematoma</td>
<td>ASCENDING 4.2 CM BY TEE (INTERNAL DIAMETER)</td>
</tr>
<tr>
<td>Penetrating ulcer</td>
<td>≥4.4–4.6 CM BY CT/MRI (EXTERNAL DIAMETER)</td>
</tr>
<tr>
<td>Marfan Syndrome</td>
<td>ANY SITE</td>
</tr>
<tr>
<td>Vascular Ehlers Danlos</td>
<td>ANY SITE</td>
</tr>
<tr>
<td>Turner Syndrome</td>
<td>ANY SITE</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>ANY SITE</td>
</tr>
<tr>
<td>Familial TAA and dissection</td>
<td>ANY SITE</td>
</tr>
<tr>
<td>Loeys Dietz syndrome</td>
<td>ANY SITE</td>
</tr>
<tr>
<td>Any patient with a growth rate of ≥0.5 cm/year</td>
<td>≥5.0–5.5 CM</td>
</tr>
<tr>
<td>Patients undergoing aortic valve repair or replacement</td>
<td>≥5.5 CM</td>
</tr>
</tbody>
</table>

Most typical location is in the descending aorta.

### Screening for associated conditions and high-risk features by history and physical examination

Patients with TAA should be screened for the presence of concomitant genetic or familial disorders which increase their individual risk of aneurysm progression or complications. This evaluation should include a thorough family history to evaluate for sudden or unexplained death and history of aneurysm or dissection. Additional historical data should include the evaluation of fevers, night sweats, arthralgias, headaches, and other constitutional symptoms that could point to a systemic inflammatory or infectious condition. Finally, risk factor analysis for coronary artery disease (CAD) and symptoms that can be attributed to CAD should be elucidated and workup for CAD pursued as appropriate. A careful physical examination to evaluate for physical features characteristic of the Marfan or Loeys Dietz syndromes, cardiac auscultation to evaluate for pathologic murmurs, and a careful peripheral vascular exam should be performed.

### Imaging

The relative advantages and disadvantages of the available techniques to evaluate the thoracic aorta are outlined in Table II. However, there are certain considerations of serial imaging in stable patients with TAA which should be addressed.

### Computed tomography scan technique

There can be significant variability in CT scan technique depending on the protocol used and the institution at which the CT scan is performed. Due to this variability, it is recommended that patients with a concern for TAA undergo a CT scan which specifically addresses the entire aorta. This is to evaluate the extent and size of the aneurysm but also detect disease in distant portions of the aorta, which may occur in up to 20% of cases. The CT scan is recommended that patients with a concern for TAA undergo a CT scan which specifically addresses the entire aorta. This is to evaluate the extent and size of the aneurysm but also detect disease in distant portions of the aorta, which may occur in up to 20% of cases.

### Magnetic resonance imaging

Magnetic resonance imaging (MRI) is an acceptable alternative to CT in stable patients with suspected thoracic aortic disease. Excellent anatomic detail and some information on valvular function are available from MRI. A comprehensive MR examination of the thoracic aorta may include many components, including black blood imaging (to evaluate aortic morphology and size and aortic wall contour) basic spin-echo sequences, non-contrast white blood imaging, and contrast-enhanced MR angiography using gadolinium based agents. The lack of ionizing radiation can make MRI a useful tool for long term surveillance of TAA, particularly in younger individuals where the accumulated potential cancer risk from repeated imaging is important. Magnetic resonance imaging reporting should sufficiently address the aorta in the same detail as outlined above with CT scanning.

### Echocardiography

If a CT or MRI has been performed that adequately assesses the aorta, it is also reasonable to obtain a transthoracic echocardiogram to screen for aortic stenosis or insufficiency, underlying bicuspid aortic valve, or other valvular disease which may alter the way in which these patients are followed and treated. In addition, evaluation of the aortic root can be suboptimal in cross-sectional imaging modalities given the asymmetry of the aortic root and artifact caused by cardiac motion. Therefore, transthoracic or transesophageal echocardiography can be a complimentary tool in which to evaluate the anatomy of the aortic root.
sufficient not sufficient alone in the initial imaging workup of suspected aortic disease.

### Medical therapy in TAA

Aggressive antihypertensive therapy is the main component of nonsurgical therapy in patients with stable thoracic aortic aneurysm. Though not based on strong evidence, smoking cessation, and possibly lipid lowering therapy and lifestyle modifications are also important aspects to medical management in these patients (Table III). Medical therapy in TAA has been most studied in the Marfan population, and the findings of these investigations can serve as a prototype for medical therapy in other TAA syndromes, given the commonalities in the proposed pathophysiologic pathways that these conditions share.37

- **β-blockers** have been shown to decrease the rate of aortic dilation, presumably due to decrease in left ventricular dp/dt and aortic shear stress.38 The use of β-blockers in patients with the Marfan syndrome carries a Class I (level of evidence B) recommendation in the recent guidelines.16 Similarly, Habashi et al and others have recently shown that angiotensin receptor blocker (ARBs) may be important in preventing aneurysm growth in the Marfan syndrome due to down-regulation of TGF-B and its effects.39,40 Angiotensin-converting enzyme inhibition may also have a beneficial effect on modifying the inflammatory milieu and in decreasing vascular smooth muscle apoptosis.41,42 Tight control of blood pressure is a Class I indication in the most recent guidelines and the specific use of ACE inhibitors, and ARBs are a Class IIa indication (level of evidence B).16 Although mostly studied in abdominal aortic aneurysm, statin therapy has the potential to mediate inflammatory markers that may contribute to aneurysm expansion and should be used to meet an low-density lipoprotein (LDL) goals as outlined by the National Cholesterol Education Program guidelines and to a goal LDL of <70 mg/dL in atherosclerotic aneurysms.43-46 However, there are no data that specifically support their use in nonatherosclerotic or inherited TAA.

### Criteria for surgical or endovascular intervention

Aortic size criteria for surgical intervention in asymptomatic patients with TAA as recently published in the TAD guidelines are illustrated in Table IV. Symptomatic patients should undergo urgent surgical evaluation as pain could represent aneurysm expansion or rupture.16 In general, all patients with an ascending aortic aneurysm of ≥5.5 cm and descending aneurysm of ≥6 cm should be offered an open surgical or endovascular procedure, depending on the location and overall surgical risk. Size criteria for intervention are smaller in those with high-risk conditions such as the Marfan syndrome, vascular ED, Turner’s syndrome, or bicuspid aortic valve.5,47-49 The data for intervening on bicuspid valve–associated aneurysm at this size is taken in part from single-center observations that evaluated patients who underwent isolated aortic valve replacement for bicuspid valve dysfunction. Those with ascending aortic dimensions of ≥4 to 4.5 cm at the time of aortic valve replacement had an increased risk of late aortic complications.50-52 The Loeys Dietz syndrome has the smallest recommended cutoff point of all genetically triggered TAA at a size of ≥4.2 cm by TEE (or >4.4-4.6 cm by CT angiography). Owing to its aggressive natural history, some have even recommended prophylactic replacement at a size of ≥4.0 cm.32

However, absolute size criteria for intervention on the aorta do not take into account natural variation in aortic size across gender and body size. Davies et al and others have proposed an aortic size indexing method that helps correct for body size.53 For example, a small woman (body surface...
Surgical technique

Open surgery

Aortic root and ascending aorta. Replacement of the portion of the aorta that is aneurysmal with a dacron or woven graft via an open approach is the most commonly performed operation for TAA in the ascending aorta. The root and ascending should be considered separately from the aortic valve in cases of proximal aortic aneurysm, that is, in cases of isolated ascending aneurysm, the valve and root should be spared if at all possible. In the Marfan population, the guidelines recommend (with level of evidence B) that patients should have excision of the sinus of Valsalva with a modified David reimplantation operation of the aortic valve when feasible, as the aortic valve itself in these patients is often normally functioning.

Aortic arch. Surgical repair of an isolated aortic arch aneurysm is challenging, carries an increased risk of stroke due to embolization of atherosclerotic material and involves the use of hypothermic circulatory arrest and cerebral perfusion techniques. In these cases, branched grafts are typically used to replace the affected portion of the arch and the great vessels.

Descending aorta. Given the morbidity associated with open procedures of the descending aorta, including spinal cord injury, endovascular therapy is becoming more common, when anatomically feasible, in management of TAA in the descending and thoracoabdominal aorta.

Endovascular therapy in thoracic aortic disease and aneurysm

Although relatively safe in high volume centers, open thoracic aorta repair still carries a significant morbidity and mortality. This is particularly true in repair of the descending aorta with some reports suggesting a postoperative paraplegia risk of nearly 15%. Although this complication may be mitigated by techniques such as distal aortic perfusion, lumbar drainage and hypothermic circulatory arrest, thoracic endovascular aortic repair (TEVAR) is taking on an increased role in the management of these patients, in particular, those who are older and with more comorbid medical conditions. Thoracic aortic stent grafting has also been used in traumatic descending aortic dissection/rupture with results that are similar to open repair. There is evidence from meta-analysis and device trials that perioperative and late-term mortality in stable patients undergoing endovascular repair of descending TAA is less than traditional open repair and associated with fewer episodes of neurologic injury.

Most thoracic endografts today are used to treat descending thoracic aneurysm disease between the left subclavian and the celiac arteries. The proximal edge of the stent graft may have to cover the left subclavian in order to provide an adequate border around the portion of the aorta excluded with the stent graft. To facilitate these cases, a carotid-subclavian bypass may be done in preparation of TEVAR. Carotid-carotid artery bypass can also be performed if the surgeon/operator feels that the left carotid also needs to be covered with the stent graft. Although not yet mainstream, it appears in some small series that TEVAR into the aortic arch may be possible with the advent of branched stent grafts.

Currently, approximately 25% of patients will be anatomically unsuitable for endovascular repair. Exclusion criteria include unfavorable “landing zones” for the stent either proximally or distally to the portion of the aorta that needs to be excluded. Unfavorable landing zones may increase the risk of endoleak and may be due to acute angulation of the aorta, heavy atherosclerotic burden, or large aneurysm extent. Poor peripheral arterial access is also a potential exclusion criteria and important to consider in patients with concomitant peripheral arterial disease.

The long-term durability of TEVAR is not yet known. However, some series have suggested that up to 20% of patients may have an endoleak, most of which can be managed percutaneously (type I). Therefore, after TEVAR, patients require ongoing follow-up and serial imaging. Computed tomography is the preferred modality to monitor for TEVAR related complications.

Surveillance imaging

Surveillance imaging is an important aspect of care in patients with asymptomatic TAA. The imaging timeline will depend on the site, etiology, and size of the aneurysm at presentation. A proposed approach to imaging these patients adapted from the 2010 guidelines on management of thoracic aortic disease is presented in Table V.

Conclusions

Thoracic aortic disease is complex and encompasses many disease states and presentations. The mortality associated with aneurysm related complications is high, and complex aortic syndromes should be managed at high-volume centers with expertise in aortic disease. Outcomes in these centers may be improved compared
to centers with lower volume. However, understanding of the disease process, medical therapy, follow-up, and indications for surgical intervention is important for all cardiovascular physicians to be familiar with in order to ensure optimal management of these patients.

Disclosures
No extramural funding was used to support this work. The authors are solely responsible for the design and conduct of this study, all study analyses, the drafting and editing of the manuscript, and its final contents.

References
## Appendix

Revised Ghent criteria for the diagnosis of Marfan syndrome

<table>
<thead>
<tr>
<th>Diagnosis of Marfan syndrome</th>
<th>Scoring system for systemic features</th>
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<tr>
<td><strong>Without family history</strong></td>
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<tr>
<td>Aortic Z-score ≥ 2 or dissection and one of the following:</td>
<td></td>
</tr>
<tr>
<td>1. Ectopic lentis</td>
<td>Wrist and thumb sign</td>
</tr>
<tr>
<td>2. Pathogenic FBN-1 mutation</td>
<td>Wrist or thumb sign</td>
</tr>
<tr>
<td>3. Systemic features: ≥ 7 points</td>
<td>Pectus carinatum</td>
</tr>
<tr>
<td>4. Ectopic lentis and FBN-1 mutation with aortic enlargement.</td>
<td>Hindfoot deformity</td>
</tr>
<tr>
<td><strong>With family history</strong></td>
<td></td>
</tr>
<tr>
<td>One of the following:</td>
<td></td>
</tr>
<tr>
<td>1. Ectopic lentis or</td>
<td>Reduced US/LS and</td>
</tr>
<tr>
<td>2. Systemic feature score ≥ 7 or reduced arm/height and increased arm/height and lack of severe scoliosis</td>
<td></td>
</tr>
<tr>
<td>3. Aortic Z-score ≥ 2 over age 20 or reduced elbow extension</td>
<td></td>
</tr>
<tr>
<td>4. Aortic Z-score ≥ 3 below age 20 or facial features†</td>
<td></td>
</tr>
</tbody>
</table>

* Aortic dimension as measured at the sinus of Valsalva. Z-Score taken from previously published data.
† Facial features (3/5 to get one point): malar hypoplasia, retr岁nathia, downsitaing palpebral fissures, enophtalmos, dolichocephaly.